# The development of a self-management education programme for people with cystic fibrosis diabetes

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# The development of a self-management education programme for people with cystic fibrosis diabetes


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# Abstract

**Background**

Cystic fibrosis diabetes (CFD) is the most common co-morbidity affecting nearly half of adults with cystic fibrosis over 30 years. Its presence has a significant impact on nutritional status, respiratory function, morbidity and mortality. Contrary to UK clinical guidelines there are currently no self-management education programmes for people with CFD. It is unclear what the barriers and facilitators are to help patients manage the additional demands of CFD, and specifically what people with CFD require to help them manage the potentially competing demands of blood glucose, nutritional status and lung function simultaneously. This study answers the research question "how can we best meet the self-management education requirements of people with CFD?"

**Methods**

This study, informed by the MRC Framework for developing and evaluating complex intervention, was conducted in two stages.

Stage 1a - a systematic search of four electronic bibliographic data bases was carried out to identify qualitative studies exploring the experience of living with and managing CFD. Meta-ethnography was used to synthesise the findings.

Stage 1b - a qualitative single occasion interview design, guided by the principles of IPA was conducted to provide insight into the self-management experiences of people with CFD and help identify skills and knowledge people with CFD require to self-manage their condition. Eight adults, from one CF centre participated.

Stage two developed and reviewed the MAGIC – Managing Abnormal Glucose in Cystic Fibrosis) programme. A stakeholder development group, consisting of 9 healthcare professional and 7 people with CFD, was established to co-design the MAGIC programme. Over 11 months, 4 workshops focusing on the content, structure and design of the MAGIC programme took place. Cognitive interviews were conducted with ten people, not previously involved in its development, to review the face validity of the MAGIC programme.

**Results**

The meta-ethnography demonstrated that individual perception of CFD directly affects its management. Many challenges in self-managing CFD were identified in the IPA study, these included: trying to balance CF and CFD, not having sufficient knowledge to inform appropriate self-management decisions and being inadequately supported by healthcare professionals. These evidences, from stage one, and expertise from the stakeholder development group informed the development of the MAGIC programme. This was developed as an on-line self-management education programme, designed for 1:1 delivery at home. It consisted of a staged approach to learning built around four modules. Its theoretical underpinnings were informed by self-efficacy theory, the self-

management framework for chronic illness and adult learning styles. The cognitive interviews identified technological problems, omissions, errors and comprehension difficulties in the MAGIC programme which were addressed before face validity was established.

**Conclusions**

This study developed the MAGIC programme, a unique self-management education programme for people with CFD. It is grounded in evidence generated from a qualitative systematic review, qualitative interviews and shared experiences of people with CFD and healthcare professional expert in the management of CFD. Recommendations for further research to assess the efficacy of the MAGIC programme include a feasibility study, a rare disease trial and the development of a core outcome set for CFD.

# Table of contents

# List of tables

# List of figures

# Author declaration

The material included in this thesis is the author's own work. No part of the thesis has been submitted in support of any academic award or qualification at another university.

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# Research Outputs

**2019**

The development of a self-management education programme for people with CFD. Oral presentation at adult and paediatric CF research away day, Royal Brompton & Harefield NHS Trust- (12th July 2019).

My PhD journey to develop a self-management education programme for people with CFD. Oral presentation at nursing and allied health professional CF education meeting, Royal Brompton & Harefield NHS Trust- (21st June 2019)

Collins, S., Jones, A., Woodward, S. & Sturt, J. (2019) P403 The experience of co-designing the MAGIC programme for people with CFD. *Journal of Cystic Fibrosis***18**, S171. Poster presentation at European CF Conference, 5-8th June 2019

Collins, S., Jones, A., Woodward, S. & Sturt, J. (2019) P302 "If I could be off them, I would" - adults with Cystic Fibrosis Diabetes experiences of corticosteroid therapy. *Journal of Cystic Fibrosis***18**, S142. Poster presentation at European CF Conference, 5-8th June 2019

PhD project management group meeting 3, 4th January 2019- presentation of the development and review of the MAGIC programme.

**2018**

The Cystic Fibrosis Diabetes (CFD) self-management triad; it is about more than just numbers. Poster presentation at Royal Brompton & Harefield NHS Trust's Nursing and Allied Health Professional Research Day, 8th November 2018.

Striking the balance between cystic fibrosis and Cystic Fibrosis Diabetes (CFD) - a meta-ethnography. Oral and poster presentation at CATO Research Symposium, Hammersmith Hospital (27th June 2018).

Collins, S., Jones, A., Woodward, S. & Sturt, J. (2018a) P160 Striking the balance between cystic fibrosis and Cystic Fibrosis Diabetes (CFD) - a meta-ethnography. *Journal of Cystic Fibrosis***17**, S104. Poster presentation at European CF Conference, 6-9th June 2018.

Collins, S., Jones, A., Woodward, S. & Sturt, J. (2018b) P161 The Cystic Fibrosis Diabetes (CFD) self-management triad; it is about more than just numbers. *Journal of Cystic Fibrosis***17**, S104. Poster presentation at European CF Conference, 6-9th June 2018.

Experiences of living with and managing CFD. Oral presentation at adult and paediatric CF research away day, Royal Brompton & Harefield NHS Trust- (27th March 2018).

PhD project management group meeting 2, 4th January 2018- presentation of IPA study.

**2017**

Striking the balance between cystic fibrosis and Cystic Fibrosis Diabetes (CFD) - a meta-ethnography. Poster presentation at Royal Brompton & Harefield NHS Trust's Nursing and Allied Health Professional Research Day, 30th October 2017. Awarded best respiratory poster award.

PhD project management group meeting 1, 20th April 2017- overview of PhD and presentation of meta-ethnography.

# List of abbreviations

| Abbreviation | |
|---|---|
| **CASP** | Critical appraisal skills programme |
| **CGMS** | Continuous glucose monitoring system |
| **CF** | Cystic fibrosis |
| **CFD** | Cystic fibrosis diabetes |
| **CFTR** | Cystic fibrosis transmembrane conductance regulator |
| **DSME** | Diabetes self-management education |
| **$FEV_1$** | Forced expiratory volume in 1 second |
| **HbA1c** | Glycated haemoglobin |
| **IPA** | Interpretative phenomenological analysis |
| **MAGIC** | Managing abnormal glucose in cystic fibrosis |
| **MRC** | Medical Research Council |
| **OGTT** | Oral glucose tolerance test |
| **PROM** | Patient reported outcome measure |

# 1 Introduction

To provide the background to this study this chapter will give: a brief review of cystic fibrosis (CF), an in-depth exploration of cystic fibrosis diabetes (CFD) and an overview of diabetes self-management education (DSME) programmes. This will be followed by an overview of my interest within this field and the outline of this thesis.

## 1.1 Background to cystic fibrosis

CF is the most common life-limiting genetic disease in the Caucasian population. It is a complex multisystem disease characterised by abnormal transport of sodium and chloride; this results in thick viscous secretions in the lungs, gastrointestinal tract, liver, pancreas and reproductive tract and elevated sodium and chloride concentrations in sweat.

## 1.2 Prevalence and survival

CF is an autosomal recessive disorder. One in 25 people in the UK carry a CF gene mutation (Cystic Fibrosis Trust 2019). CF occurs in approximately one in every 2500 live births (Dodge et al. 2007) and currently affects 10,461 people in the UK (Cystic Fibrosis Trust 2018a). In the 1930's, when CF was first described, 70% of babies died within the first year of life (Andersen 1938). Of those born in the 1970's over half died before they reached mid-late teens (Dodge et al. 2007). The median predicted age of survival, for a child born today with CF, is 47 years (Cystic Fibrosis Trust 2018a). Improved survival is attributed to many factors which include: specialist CF centre multidisciplinary care, improved nutritional management, routine airway clearance techniques, improved antibiotic therapy (Plant et al. 2013) and newborn screening (Dijk et al. 2011). Improvements in survival has resulted in the development of co-morbidities which increase the complexities of treating and managing CF.

## 1.3 The underlying genetic defect

The CF gene was first identified in 1989 (Kerem et al. 1989); it codes for a protein called the cystic fibrosis transmembrane conductance regulator (CFTR). This protein

acts as a chloride channel and is responsible for the movement of chloride ions across the apical membrane of epithelial cells (Ratjen 2009). Chloride ions affect the amount of water moved in and out of cells. CFTR is widely found in epithelial cells of the airways, salivary glands, sweat glands, pancreas, intestines and reproductive tract and this is where the major clinical manifestations are exhibited (Davis 2006). CF causing gene mutations lead to defective or absent CFTR, which in turn results in elevated concentrations of sodium and chloride in sweat and viscous secretions in organs such as the lungs, gastrointestinal tract and the male reproductive system.

More than 2000 CFTR variants have been identified. Six classes of CFTR mutations have been recognised (table 1); classification is based upon the impact on CFTR protein function. F508del, a class II mutation, is the most common CFTR mutation; approximately 90.9% of the UK CF population have at least one copy of the F508del mutation (Cystic Fibrosis Trust 2018a). Class II mutations result in CFTR protein that is not folded or processed correctly. The relationship between different gene mutations and clinical phenotype is complex and often result in variable levels of disease severity, this is even seen between siblings with the same genotype (O'Sullivan & Freedman 2009). Environmental factors and other genetic components such as CF modifiers have also been proposed in playing a role in the clinical outcome of people with CF (Zielenski 2000).

**Table 1 Classes of CFTR mutation**

| Class | CFTR defect |
|---|---|
| **I** | No functional CFTR protein |
| **II** | CFTR trafficking defect |
| **III** | Defective channel regulation |
| **IV** | Decreased channel conductance |
| **V** | Reduced synthesis of CFTR |
| **VI** | Decreased CFTR stability |

## 1.4 Diagnosis

Since 2007 all babies born in the UK are screened for CF as part of the Newborn Screening Programme. This leads to earlier diagnosis which has been shown to have a

positive effect on nutritional status (Farrell et al. 1997, Leung et al. 2017), respiratory outcomes (Cystic Fibrosis Foundation et al. 2009) and survival (Dijk et al. 2011). The majority of people with CF are therefore now diagnosed in infancy but those born before the introduction of newborn screening may be diagnosed in later life. Symptoms of CF include; faltering growth/ undernutrition, protracted diarrhoea, recurrent respiratory infections, pancreatitis and male infertility. The diagnosis of CF later in life as a result of clinical manifestations is confirmed by sweat testing, CF genotyping and measures of CFTR function such as the nasal potential difference test (Farrell et al. 2008).

## 1.5 Clinical manifestations

### 1.5.1 Respiratory tract

In the CF respiratory tract mucus secretions contain less water and are therefore viscous and more difficult to clear. The presence of viscous mucus in the lungs increases the risk of colonisation by pathogens and leaves the lungs susceptible to infections and inflammation. Recurrent respiratory infections lead to progressive lung damage and loss of lung function which can ultimately result in death from respiratory failure.

### 1.5.2 Gastrointestinal tract

CF has a major effect on the gastrointestinal system and gastrointestinal complications include: pancreatic exocrine insufficiency, gastro-oesophageal reflux disease and distal intestinal obstructive syndrome.

Pancreatic exocrine insufficiency is the most common gastrointestinal complication, affecting between 80 to 90% of people with CF (Somaraju & Solis-Moya 2016). Damage to the pancreas begins in utero and is progressive; pancreatic cells are replaced with fibrous scar tissue as a result of tissue damage caused by the deposits of dried up secretions. This inhibits pancreatic function causing a reduction in the production of pancreatic enzymes. Pancreatic enzymes play an important role in the digestion of macronutrients, predominately proteins, fats and carbohydrates. When the quantity of pancreatic enzymes secreted in response to a meal is insufficient to maintain normal

digestive processes pancreatic exocrine insufficiency occurs. Fat maldigestion and malabsorption are the major clinical manifestation of pancreatic exocrine insufficiency, if left untreated they will result in sub-optimal nutritional status and deficiency of fat soluble vitamins (Cystic Fibrosis Trust 2016).

Pancreatic enzyme replacement therapy is used to treat pancreatic exocrine insufficiency; it is prescribed with all fat and protein containing meals and snacks. The aims of treatment are to achieve adequate digestion of all nutrients thereby controlling the signs and symptoms of malabsorption. This is essential in order to achieve sufficient growth and development, preserve normal nutritional status and obtain adequate fat-soluble vitamin levels.

### 1.5.3 Liver disease

The majority of hepatobiliary complications seen in people with CF present in childhood or early adolescence. The clinical presentation of CF liver disease is variable and ranges from asymptomatic elevation in serum liver enzymes to cirrhosis. Liver steatosis, which has been shown to have little clinical impact, is the most common finding, affecting 23-75 percent of people with CF (Lewindon & Ramm 2011, Parisi et al. 2013). Cirrhosis is irreversible; in 5-15 percent of cases focal biliary cirrhosis leads to multilobular biliary cirrhosis and portal hypertension (Parisi et al. 2013). Liver failure is uncommon, mainly affecting older children and adults (Debray et al. 2011).

The aim of treatment of cystic fibrosis liver disease is to prevent liver damage and complications associated with portal hypertension and cirrhosis. Ursodeoxycholic acid is widely used to treat people with cystic fibrosis liver disease (Moyer & Balistreri 2009). Liver transplantation is an option for some people with end-stage cystic fibrosis liver disease.

### 1.5.4 Reproductive system

Approximately 98% of men with CF are infertile due to the blockage or absence of the vas deferens. Advances in fertility treatments involving sperm aspiration and intra-cytoplasmic sperm injection (ICSI) has led to many men with CF fathering children (Lyon & Bilton 2002). Women with CF may well have normal fertility, however the risks

of pregnancy upon maternal health including the unpredictable impact of pregnancy upon lung function and challenges in maintaining adequate nutrition should be considered prior to conception (Edenborough et al. 2008).

### 1.5.5 Bone disease

Low bone mineral density is a common complication seen in children and adults with CF. Its aetiology is multifactorial and risk factors include; deficiencies of vitamin D, vitamin K and calcium, CFTR dysfunction, poor nutritional status, glucocorticoid use, hypogonadism, respiratory exacerbations and low physical activity levels (Sermet-Gaudelus et al. 2011). Prevention and detection of low bone mineral density are an important part of routine CF care because of the associated risk of fragility fractures.

## 1.6 Management of cystic fibrosis

### 1.6.1 Multidisciplinary approach to care

All people with CF receive their care from a Specialist CF Centre, this care should be delivered by a specialist multidisciplinary team of trained and experienced healthcare professionals (UK Cystic Fibrosis Trust Standards of Care Working Group 2011, NICE 2017). The multidisciplinary team should contain specialist consultants, clinical nurse specialists, dietitians, physiotherapists, clinical psychologists, social workers, pharmacists and administrative support.

### 1.6.2 Medical therapy

There is no cure for CF; medical management aims to maintain health by helping to control symptoms of the disease. Medical management of respiratory disease includes: the use of oral, inhaled and intravenous antibiotics to help to treat or control persistent infections, bronchodilators to open up the airways, corticosteroids to reduce inflammation in the airways and mucolytics such as rhDNASe which decreases the viscosity of airway secretions and assists sputum clearance.

### 1.6.3 Physiotherapy

A CF specialist physiotherapist will provide individualised advice, this includes appropriate airway clearance techniques and exercise. Daily airway clearance is

required to aid the removal of bronchial secretions; this is often in conjunction with the use of mucolytic agents. Keeping the airways clear of secretions helps reduce lung inflammation and infections and improves lung function (Association of Chartered Physiotherapists in Cystic Fibrosis 2017).

### 1.6.4 Nutrition

Progressive respiratory disease is the major cause of mortality in people with CF however, good nutritional status has been associated with improved survival (Corey et al. 1988, Stallings et al. 2008). Intensive nutritional management involving a high-energy, high-fat diet and pancreatic enzymes has been the mainstay of treatment for people with CF over the past 35 years and nutritional failure is no longer accepted as a consequence of CF (Collins 2018).

There is a wide variation in energy requirements between people with CF, however requirements are generally quoted as being 110-200 percent of those required by healthy individuals of the same age and gender (Stallings et al. 2008). Energy requirement will vary according to nutritional and clinical status, age, gender, presence of infections and control of malabsorption. People with CF who are pancreatic sufficient tend to have lower energy requirements and fewer nutritional problems than those who are pancreatic insufficient. For some people with CF, despite following a high-fat, high-energy diet, nutrition will be a problem and supplementary nutritional support may be required. This will usually follow a staged approach of first trying oral nutritional supplements, only progressing to enteral tube feeding if nutritional requirements are not achieved (Cystic Fibrosis Trust 2016).

Improvements in survival and nutritional status in people with CF have contributed to overweight and obesity becoming a more frequent problem (Panagopoulou et al. 2014, Hanna & Weiner 2015). In the UK approximately 17% of adults are overweight and 4% are obese (Cystic Fibrosis Trust 2016). Obesity is not just seen in people with CF who are older and pancreatic sufficient (Stephenson et al. 2013) it is now an emerging problem in children and people with CF who are pancreatic insufficient (Hanna & Weiner 2015).

A CF specialist dietitian is an integral part of the multi-disciplinary team providing on-going nutritional assessment and appropriate dietary advice. This will facilitate normal growth/ development, optimise nutritional status and support long-term health and well-being.

### 1.6.5 Psychosocial care

CF is a progressive life-limiting disease, which impacts on all aspects of day-to-day life. Treatment regimens for people with CF are complex, with a high burden of treatment; routine CF care can take two-three hours per day (Sawicki et al. 2009). People with CF and their carers can often experience difficulties managing this chronic disease, its treatment demands and the impact it has upon their lives. People with CF should have access to psychosocial support from clinical psychologists and social workers as part of their routine CF management (UK Cystic Fibrosis Trust Standards of Care Working Group 2011, NICE 2017).

### 1.6.6 Cross infection

Cross-infection with different respiratory pathogens between people with CF can be detrimental (Whiteford et al. 1995). For this reason, people with CF are advised not to meet face to face. Group-based based activities involving direct contact are therefore prohibited. CF centres are advised to follow infection control guidelines to segregate patients in wards and outpatient clinics (UK Cystic Fibrosis Trust Standards of Care Working Group 2011, NICE 2017). The loss of peer support between people with CF is a negative consequence of segregation (Quittner et al. 2016b). Social media and online forums are used amongst people with CF to offer peer support, however a recent study highlighted that despite the use of electronic communications adolescents with CF were still meeting in person (Helms et al. 2015). This implies the need for heightened education amongst this vulnerable age-group of the CF population regarding the risks of cross-infection.

## 1.7 Future CF care

### 1.7.1 Ageing CF population

Predictions indicate that the number of people with CF in the United Kingdom will increase by 54% from 2010 to 2025 due to improvements in survival. There will be a 79% increase in the adult population and 25% increase in the paediatric population (Burgel et al. 2015). This will have a significant impact on the provision of CF care, with the need to develop more CF centres, particularly for adults, to be able to continue to provide high-standard CF care.

### 1.7.2 Correcting the Underlying CFTR Protein Dysfunction

Treating the basic defect underlying the CF phenotype is an approach to address many of the on-going treatment challenges people with CF experience. The first drugs developed to treat the underlying causes of CF are CFTR potentiators, these work by improving CFTR channel function, and correctors that work by improving the number of CFTR channels (Burgener & Moss 2018). As different mutations cause different defects in CFTR protein, the medications developed only are effective in people with specific mutations. Therefore, these recent advancements have made addressing the basic CF defect realistic for the first time, for some.

Recently a study with Ivacaftor (a CFTR potentiator), highlighted that there may be a period in early life where some repair of pancreatic exocrine function may be possible (Davies et al. 2015). Further research is needed with regards to this and to also ascertain the potential impact of CFTR potentiators on pancreatic endocrine function.

### 1.7.3 Co-morbidities

With improved life expectancy, people with CF are at increased risk for co-morbid conditions that may have an impact upon mortality and quality of life. Cystic fibrosis diabetes (CFD) is the most common co-morbidity in people with CF. UK CF registry data demonstrates a much higher prevalence of CFD (33.8%) in adults compared to other co-morbidities such as: gastro-oesophageal reflux disease (19.7%), osteopenia (18.2%), liver disease (16.2%), osteoporosis (7.3%) and distal intestinal obstructive syndrome (7.3%) (Cystic Fibrosis Trust 2018a).

## 1.8 Background to cystic fibrosis diabetes

Since the late 1990s the term cystic fibrosis related diabetes has been used to describe diabetes in people with CF. Prior to this it was referred to as diabetes of cystic fibrosis or cystic fibrosis diabetes mellitus. To reflect the findings and recommendations of this study the term cystic fibrosis diabetes will be used throughout this thesis. The reasons for this will be explored in chapter three.

People with CF and pancreatic insufficiency experience a gradual reduction in insulin production over time and, this causes the higher prevalence of CFD seen in the adult CF population. CFD is a serious co-morbidity with significant impact on morbidity and mortality.

### 1.8.1 Prevalence

The prevalence of CFD increases with age. Data from the USA shows it affects 2% of children, 19% of adolescents, 40% of adults in their 20s and 45-50% of adults $\geq$30 years (Moran 2009). In the UK data from the CF Trust's Registry Report shows 10.2% of children aged 10-15 years and 33.8% of the adults 16, or older, are on CFD treatment (Cystic Fibrosis Trust 2018a). The CF Trust Registry only records people diagnosed with CFD who are on CFD treatment. Therefore, anyone diagnosed with CFD but not on treatment, such as those on dietary modifications, are not included in this registry data.

CF genotype has an association with the prevalence of CFD. Lewis et al. (2015) found several associations between CF genetic and CFD. In people with CF and severe genotypes (no functional CFTR protein expressed, pancreatic insufficiency) prevalence of CFD is higher than those with mild genotype (mild CFTR dysfunction, pancreatic sufficiency) - 60% vs 14% $p<0.0001$). A gender variation was also seen with 66% females with a severe genotype compared to 54% males developing CFD. There was a significant increase in the prevalence of CFD with age - 19 of 23 women (83%) compared to 18 of 28 men (64%), aged 40-49 with severe genotype had CFD. While these numbers are small, they are an important consideration with increased

predictions of survival in the CF population and thus growing numbers of adults in this older age group.

### 1.8.2 Pathophysiology

CFD has a complex pathophysiology. CFD shows features of both type 1 and type 2 diabetes (table 2) but it is a distinct clinical entity with important pathological differences (Moran et al. 2018). The primary cause of CFD is insulin deficiency, but factors distinctive to CF such as undernutrition, malabsorption, increased energy expenditure, glucagon deficiency, liver dysfunction, chronic and acute infections influence glucose metabolism (Dyce et al. 2015, Moran et al. 2018). Glucose tolerance may therefore fluctuate over time, as these factors are not static.

| | Type 1 | Type 2 | CFD |
|---|---|---|---|
| **Cause** | Autoimmune destruction of β cells & near complete insulin deficiency | Insulin resistance and effect of glucose on pancreatic function | Severe, not complete insulin deficiency<br>Insulin sensitivity reduced during acute illness |
| **Onset** | Usually acute | Insidious | Insidious |
| **Peak age of onset** | Children- late 30s * | Adult[+] | 18-24 years |
| **Usual nutritional status** | Generally normal weight | Obese | Generally normal- underweight |
| **Diagnosis and screening** | Clinical features<br>Elevated random glucose levels | Clinical features<br>Oral glucose tolerance test, elevated fasting plasma glucose | Clinical features are rare<br>Oral glucose tolerance test & continuous glucose monitoring<br>Fasting plasma glucose unhelpful |
| **Usual treatment** | Insulin | Diet, oral agents, insulin | Insulin |
| **Treatment rationale** | Reduction in osmotic effects of high glucose.<br><br>Reduction in long-term complication risks | Reduction in osmotic effects of high glucose<br><br>Reduction in long-term complication risks | Reduction in the impact on high glucose on lung function, infection risk and nutritional status<br>Reduction in long-term complication risks |
| **Dietary management** | Healthy balanced diet<br><br>Reduction simple carbohydrates | Healthy balanced diet<br><br>Weight reduction<br>Reduction simple carbohydrates | Usual energy intake to meet requirements (100-200% of normal)<br>Optimisation of nutritional status<br>Modification of simple carbohydrates |
| **Complications** | Risk of micro- and macrovascular complications | Risk of micro- and macrovascular complications | Significant effects of hyperglycaemia on nutritional and respiratory status<br>Risk of microvascular complications<br>Macrovascular complications are rare |
| **Ketosis risk** | Yes | Unusual | Rare |
| **Cause of death** | Cardiovascular | Cardiovascular | Respiratory |

**Table 2 Comparison of features of different types of diabetes**

* Type 1 diabetes used to be regarded as a disease of children and adolescents however approximately fifty percent of newly diagnoses cases of type 1 diabetes are now in people greater than 18 years old (JDRF 2018).

[+]More cases of type 2 diabetes are being seen in young people, currently in the UK 1.9% of those < 19 years with diabetes have type 2 (Diabetes UK 2016a)

#### 1.8.2.1 Insulin deficiency

The primary cause of CFD is insulin deficiency. There is destruction of the pancreatic tissue, caused by thick mucus secretions and obstructive damage, with accompanying fibrosis and disorganisation of the islet cells, which gradually result in a reduction in insulin production (Moran et al. 2010a). The reduction in β cell numbers in CFD is not related to autoimmune disease as is the case in type 1 diabetes (Bridges et al. 2018). People with CFD do not have complete insulin deficiency as seen in type 1 diabetes, they maintain some residual insulin production (Moran et al. 2018). Initially a delay and reduction in peak insulin production is seen and the secretion of basal insulin is often preserved (Perano et al. 2014).

Pancreatic destruction is not the only reason for glycaemic abnormalities in CF. Animal models of CFD suggest a functional deficiency in islet cells plays a part (Moheet & Moran 2017). Newborn CF pigs have been shown to have abnormal insulin secretion and reduced responsiveness to hyperglycaemia, which was not caused by reduced islet cell mass (Uc et al. 2015). Similar observations have been seen in the CF ferret which from birth demonstrated abnormal glucose tolerance (Olivier et al. 2012). Glycaemic abnormalities have also been seen in infants and children with CF, where insufficient insulin secretion has been proposed as the cause (Yi et al. 2016).

#### 1.8.2.2 Insulin resistance

In people with CF, insulin sensitivity is usually normal (Moran et al. 1994). Where there is another reason for insulin resistance such as acute respiratory infection, corticosteroid treatment or immunosuppressive therapy reduced insulin sensitivity may cause hyperglycaemia (Bridges et al. 2018). Insulin resistance is not as important as insulin deficiency in the pathogenesis of CFD, but it will vary throughout the clinical course of CF and therefore needs consideration.

#### 1.8.2.3 Impairment in glucagon secretion

Glycaemic abnormalities in people with CF can also be attributed to impairment of glucagon secretion due to total islet destruction (Moran et al. 2018). Like insulin, glucagon is a hormone produced in the pancreas, but its role is to raise blood glucose levels.

In people with CFD, unlike those with type 1 diabetes, the risk of ketoacidosis is rare. This is because of the impairment in glucagon secretion and/or maintenance of some residual insulin secretion (Moran et al. 2018).

#### 1.8.2.4 CFTR protein

CFD mainly occurs in people with CFTR mutations associated with more severe disease including pancreatic exocrine insufficiency (Moran et al. 2018). The CFTR protein may have a role in glycaemic abnormalities in CF (Prentice et al. 2016) because CFTR has a role in insulin secretion and β cell function (Osorio 2014). From birth, CF ferrets have demonstrated abnormal insulin secretion signifying, that CFTR might play a role in insulin secretion (Olivier et al. 2012).

### 1.8.3 Clinical manifestations

In people with CF, a decline in body weight and lung function has been seen in the early stages of abnormal glucose metabolism, prior to the diagnosis of CFD (Lanng et al. 1992, Brodsky et al. 2011, Hameed et al. 2015). Insulin deficiency is associated with a catabolic state, which may be a potential mechanism for decline in both respiratory and nutritional status (Moheet & Moran 2017). It is less clear whether hyperglycaemia or insulin deficiency have a causal relationship with poor outcomes, such as lower lung function, sub-optimal nutritional status or reduced survival, or are simply markers of more advanced or complicated disease (Jones & Sainsbury 2016). Due to the complexities in the clinical manifestation of CFD it is not clear if sicker people with CF get diabetes or if diabetes makes people with CF sicker.

#### 1.8.3.1 Impact on survival

CFD is associated with increased mortality (Finkelstein et al. 1988, Chamnan et al. 2010, Lewis et al. 2015). Lewis et al. (2015) recently demonstrated that despite earlier identification and treatment of CFD aged-adjusted mortality remains 3.5 times greater in people with CFD compared to those without (Lewis et al. 2015). Being female is typically associated with worse survival in CF (Corriveau et al. 2018). However, CFD increased the risk of death in both men and women, the protective effects of the male sex seen in CF men without diabetes was no longer apparent (Lewis et al. 2015). It is unclear what is the cause of mortality in people with CFD because hyperglycaemia has

been shown to have a variable impact on survival (Adler et al. 2011, Lewis et al. 2015). Hyperglycaemia, as defined as HbA1c >6.5%, has been associated with a threefold increase risk of death in people with CFD (Adler et al. 2011). Conversely HbA1c and duration of CFD were found to be similar between aged-matched survivors and those who died during their 30s and 40s (Lewis et al. 2015). This suggests that even well-controlled diabetes adds an additional inflammatory burden that together with CF-related respiratory inflammation creates a greater risk of death for these individuals (Lewis et al. 2015).

#### 1.8.3.2 Impact on lung disease

In a cross-sectional study of >7500 people with CF a lower forced expiratory volume in 1 second ($FEV_1$) was observed in all age groups with CFD (Koch et al. 2001). The mean $FEV_1$ was 72%, at all ages, in those without diabetes compared to 52% in those with CFD. Moran et al. (2009a) also demonstrated that lung function remains lowered in people with CFD, this was despite earlier diagnosis and initiation of insulin.

People with CFD also experience a higher rate in decline in lung function ($FEV_1$) compared to those with normal glucose tolerance (Milla et al. 2000). Lung function decline has been reported as being nearly twice as high, with an average yearly rate in $FEV_1$ decline of 3.2% in those with CFD compared to 1.8% in those without (Mohan et al. 2008). Despite earlier diagnosis and insulin treatment this effect is only partially reversed with CFD treatment (Rolon et al. 2001, Dobson et al. 2002). Mohan et al. (2008) demonstrated significant improvement in lung function at 3 months and one year following insulin initiation however, this was not sustained. At 2 and 3 years post-insulin initiation the rate of lung function decline observed was similar to that of pre-treatment ( -3.2% $FEV_1$ per year).

The mechanism that links CFD with inflammation and infection is not fully understood; *in vitro* studies have demonstrated that increased airways surface liquid glucose concentration is associated with an increased growth of *Staphylococcus aureus* and *Pseudomonas aeruginosa* in the airways (Brennan et al. 2007, Bradbury et al. 2009). Blood glucose levels of > 8mmol/l are associated with elevated airways glucose concentrations in people with CF, thus contributing to higher concentrations of

airways bacteria (Brennan et al. 2007). This is a serious concern for people with CF. Lung damage is a result of the vicious cycle between infection and inflammation. Mucus blocks the airways, allowing bacteria to grow and cause infections. An increase production of mucus is the result of infection, this causes inflammation which damage cilia in the airways and leads to difficulties clearing mucus secretions from the lungs. Therefore, hyperglycaemia causes elevated glucose in the airways, which in turn promotes the growth of airway bacteria and contributes to respiratory infections and lung function decline (Hameed et al. 2015).

#### 1.8.3.3 Impact on nutritional status

Insulin is an anabolic hormone essential to the body for the metabolism of protein, fats and carbohydrates. Its primary role is to promote the absorption of carbohydrates (particularly glucose) from the blood into fat, skeletal muscle and the liver. Hence a deficiency of insulin would lead to a worsening of nutritional status because: the body would not be able to turn ingested glucose into energy, glucose released by the liver through the process of glycogenolysis would not be converted to energy and the body would start breaking down its stores of protein and fat as an alternative source of energy. Insulin deficiency in CF is insidious, therefore the slow gradual loss of insulin over time causes weight and lean body mass loss (Bridges et al. 2018). It is not unusual to observe a decline in BMI for several years prior to the diagnosis of CFD (Finkelstein et al. 1988, Lanng et al. 1992, Hameed et al. 2010). Unintentional weight loss is not uncommon in type 1 diabetes presentation (Diabetes UK 2019) however in CFD it presents much more slowly.

Hyperglycaemia will also promote glycosuria and hence the loss of energy in urine. This can also contribute to difficulties in maintaining weight and/or weight loss.

#### 1.8.3.4 Microvascular and macrovascular complications

With improvements in survival, the risk of diabetic complications in people with CFD requires attention. Duration of diabetes and glycaemic control are two influential factors in the risk of developing microvascular complications (Andersen et al. 2006, Schwarzenberg et al. 2007). A study of people who had CFD and fasting hyperglycaemia, for more than 10 years found the rates of microvascular

complications to be less than that seen in people with type 1 or type 2 diabetes, with microalbuminuria, retinopathy, neuropathy and autonomic gastropathy affecting 14% 16%, 55% and 50% respectively (Moran et al. 2010b). The presence of some residual insulin secretion in people with CFD and absence of metabolic abnormalities seen in type 2 diabetes may contribute to this lower prevalence (Moheet & Moran 2017). In another study nephropathy and neuropathy rates were reported as similar to those with type 1 diabetes, however a lower rate of retinopathy was seen (van den Berg et al. 2008). Other risk factors associated with retinopathy in this type 1 population such as smoking, higher HbA1c, cholesterol and BMI may have been additional contributory factors to developing retinopathy. Retinopathy has also been seen in three individuals with abnormal glucose tolerance who do not meet the criteria for CFD (Gilchrist et al. 2015).

Routine annual examination for neuropathy and retinopathy along with urine screening for microalbuminuria are recommended for people with CFD (Cystic Fibrosis Trust 2004, Moran et al. 2010b, Middleton et al. 2014), however this guidance needs revision in light of cases of retinopathy developing in abnormal glucose tolerance (Gilchrist et al. 2015). Macrovascular complications are rare in people with CFD due to low cardiovascular risks factors seen in this population (Moheet & Moran 2017).

### 1.8.4 Diagnosis

The development of CFD tends to be insidious due to slow and progressive deficiency of insulin and hence gradual deterioration in glucose tolerance. Few people with CF have completely 'normal' glucose tolerance and CFD is viewed as part of a continuum of glucose tolerance abnormalities (Figure 1) (Moran et al. 2010a). Glucose tolerance can vary due to the clinical variability in CF e.g. respiratory exacerbations, transient use of corticosteroid treatment, periods of increased glucose loads such as the use of enteral tube feeding (Moran et al. 2018). In clinical practice it is not unusual for people with CF to only require insulin treatment during periods of illness, corticosteroid use or enteral tube feeding.

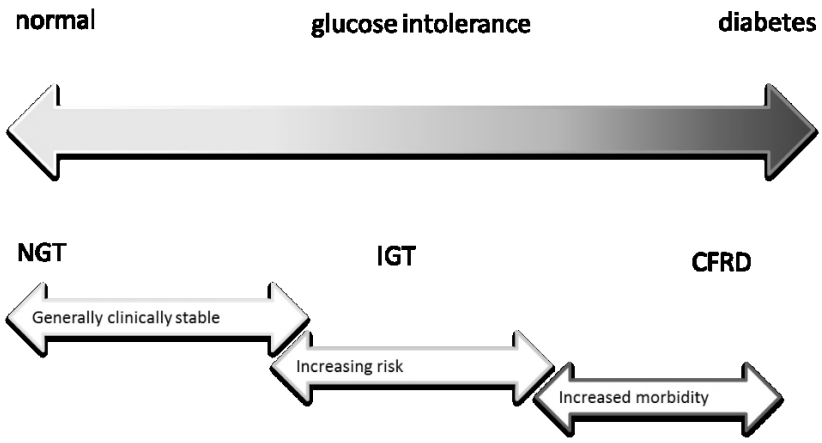


**Figure 1 The clinical spectrum of glucose tolerance in CF**

The diagnosis of CFD can be difficult to make because the majority of people with CFD do not show any obvious symptoms (table 3) at the time of diagnosis (Moran et al. 2018). The median age (interquartile range) of diagnosis of CFD is 18.7 years (15.5-25) (Konrad et al. 2013). Early intervention can protect against a decline in lung function and weight loss therefore CFD screening programmes are recommended as standard practice (Cystic Fibrosis Trust 2004, Middleton et al. 2014, Moran et al. 2018).

**Table 3 Symptoms of CFD**

| Symptoms of CFD |
|---|
| Unexplained polyuria or polydipsia |
| Blurred vision |
| Inability to maintain or gain weight despite nutritional intervention |
| Inadequate growth velocity |
| Unexplained decline in lung function |
| Increased frequency of respiratory exacerbations |
| Delayed puberty |

### 1.8.5 Screening for CFD

The importance of monitoring blood glucose levels due to the clinical effects of CFD is well recognised, with annual screening for CFD recommended from 10 years of age (Cystic Fibrosis Trust 2004, Middleton et al. 2014, Turck et al. 2016, Moran et al. 2018).

### 1.8.5.1 Oral glucose tolerance test

The oral glucose tolerance test (OGTT) is used to diagnose diabetes by measuring the body's response to glucose. Following an overnight fast (8-14 hours) a fasting blood glucose sample is taken. Patients are then required to drink a glucose solution containing up to 75g of glucose. A further blood sample is taken two hours after the test load. Table 4 identifies the WHO criteria for the diagnosis of diabetes (WHO 2018).

**Table 4 WHO Criteria for the diagnosis of diabetes (WHO 2018)**

| Diagnostic category | Diagnostic criteria |
|---|---|
| **Diabetes** | Fasting plasma glucose ≥7.0mmol/l<br>or<br>2–h plasma glucose ≥11.1mmol/l |
| **Impaired glucose tolerance** | Fasting plasma glucose <7.0mmol/l<br>and<br>2–h plasma glucose ≥7.8 and ≤11.1mmol |
| **Impaired fasting glucose** | Fasting plasma glucose 6.1 to 6.9mmol/l<br>and (if measured)<br>2-h plasma glucose <7.8mmol/l |

The use of OGTT as a screening tool for CFD has received much criticism for many reasons. Firstly, the OGTT is designed to diagnose diabetes. It may not identify the early stages of dysglycaemia seen in CFD, which start to impact weight and lung function before the diagnostic criteria for diabetes are met. This highlights important questions about early dysglycaemia in CF and the methods used to detect this (Dobson et al. 2004, Hameed et al. 2011, Schmid et al. 2014). Secondly, the relevance of the 2-hour OGTT has been questioned. The 60- minute OGTT has been associated with indicators of clinical deterioration in adults with CF and may therefore be a more appropriate measure to use (Coriati et al. 2016). Similarly sampling blood at 30 or 60 minutes has been shown to identify earlier glucose abnormalities to alert insulin deficiency before the onset of CFD (Dobson et al. 2004, Hameed et al. 2011). Thirdly, the cut-off values from OGTT determining CFD were taken from type 2 diabetes (table 4) in which they were used to predict microvascular complications, where in CFD

treatment is initiated to prevent respiratory and nutritional decline (Bridges et al. 2018). Finally, wide variations in glucose tolerance are observed in people with CF, particularly due to the effects of insulin resistance during infections therefore OGTT results will be subjected to wide variability throughout the clinical course of CF (Dyce et al. 2015).

Due to low sensitivity random glucose, urinary glucose and HbA1c are not recommended as screening tools for CFD (Moran et al. 2010b). HbA1c is unreliable in people with CF because it can be artificially low due to increased red blood cell turnover (Moran et al. 2010a) or high in those with iron deficiency (Bridges et al. 2018). High blood glucose levels may also be intermittent in the early stages of CFD and not significant enough to elevate HbA1c (Prentice et al. 2016). HbA1c $\geq$ 5.8% (40mmol/mol/) has been proposed as a screening tool to predict the need for an OGTT in an adult CF clinic (Burgess et al. 2016) although this practice has been met with criticism for still not being sensitive enough to identify early glucose abnormalities (Widger et al. 2016).

Following a positive OGTT a period of serial blood glucose monitoring or continuous glucose monitoring is needed to identify when elevations in blood glucose levels occur and to guide treatment decisions. The question when to initiate treatment for dysglycaemia is not clear cut. Bridges et al. (2018) suggest that people who meet the diagnostic criteria for diabetes should be treated and in those with impaired glucose tolerance on OGTT or post-prandial blood glucose >8mmol/l treatment should be considered if weight or lung function is declining.

#### 1.8.5.2 Continuous Glucose Monitoring Systems

Continuous glucose monitoring systems (CGMS) are increasingly being used in CF centres, in addition to or instead of OGTT, to assist in the early identification of abnormal glucose metabolism and to inform treatment decisions (O'Riordan et al. 2009, Dyce et al. 2015). It is however not licenced as a tool to diagnose any type of diabetes (Prentice et al. 2016). CGMS involves the insertion of a small probe (figure 2), into the subcutaneous tissue of the abdomen, which is attached to a transmitter allowing the measurement of interstitial glucose. The CGMS averages the glucose

levels approximately every five minutes and can be worn for up to ten days. It can be inserted quickly and easily in the clinical setting and has the additional benefit of allowing individuals to be at home following their usual routines and diet. CGMS has been shown to correlate with plasma glucose levels in people with CF (Dobson et al. 2003, O'Riordan et al. 2009). Some CGMS may require blood glucose measurements (via finger-prinks) for calibration (Dexcom 2018, Medtronic 2018). A useful addition to CGMS are the visual reports obtained (figure 3); in clinical practice these can assist with treatment initiation decisions, dietary changes, treatment modifications and patient education.

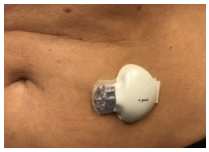

**Figure 2 IPro2 sensor**

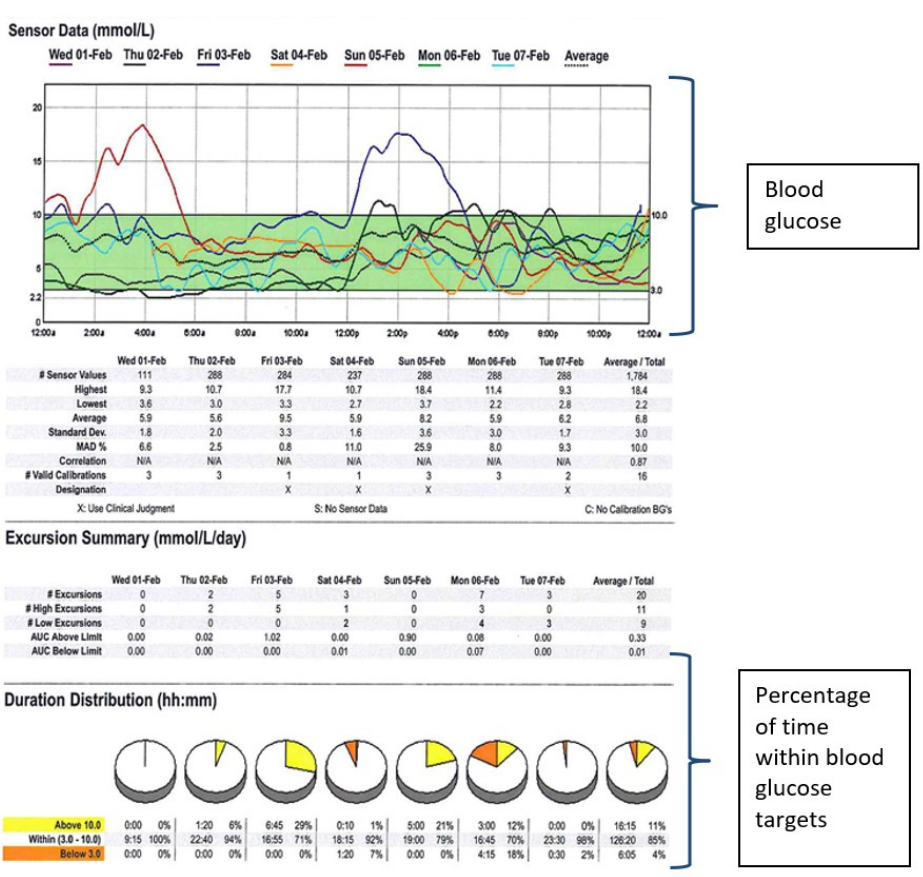


**Figure 3 Example of CGMS data report**

One of the significant factors for initiating treatment of dysglycaemia is the impact on nutrition. CGMS has been used to highlight children at risk of nutritional decline, where a CGMS value greater than 7.8mmol/l for over 4.5% of the test was predicative of a decline in weight standard deviation scores (Hameed et al. 2010). However, there are no definitive criteria when to start insulin based on CGMS. One paediatric CF centre has modified the findings from Hameed's research to guide its treatment decisions- table 5 (RBHFT 2017). However, in clinical practice treatment decisions tend to be made by CFD teams on an individual basis (Cystic Fibrosis Trust 2004, Middleton et al. 2014, Moran et al. 2018).

**Table 5 CFD management strategy based on CGMS results (RBHFT 2017)**

| Diagnostic category | CGM values | Treatment |
|---|---|---|
| **CFD** | 2 x peaks >11.1mmol/l<br>and<br>>10% of time >7.8 | Start insulin |
| **Impaired glucose tolerance** | No more than 1 peak >11.1mmol/l<br>and/or<br>>10% of time >7.8mmol/l | Consider insulin<br>Repeat CGMS in 6 months |
| **Indeterminate glucose homeostasis** | 4.5-10% of time >7.8mmol/l or hypoglycaemia | Close monitoring<br>Dietary modification for hypoglycaemia<br>Repeat CGMS in 12 months |
| **Normal** | No peaks >11.1mmol/l<br>and<br><4.5% of time >7.8mmol/l | None<br>Repeat CGMS when indicated |

### 1.8.6 Management

In type 1 or type 2 diabetes the rationale for treating hyperglycaemia is to minimise osmotic symptoms and reduce the risk of long-term diabetic complications (Bridges et al. 2018). Treating CFD is not just about correcting hyperglycaemia, its primary focus is on preserving or improving clinical and nutritional status and hence influencing survival (Moheet & Moran 2017). The aims of management of CFD are therefore: to maintain optimal growth, nutritional status and respiratory function, to achieve good control of blood glucose levels and to avoid long-term diabetic complications (Cystic Fibrosis Trust 2016). The optimal treatment of CFD is controversial with no consensus

as to when and how to treat. This reflects the complexities of CFD and the uniqueness of the individual. What is clear, however, are the benefits of early treatment.

#### 1.8.6.1 Insulin

All clinical guidelines recommend insulin therapy as first line treatment because the primary cause of CFD is insulin deficiency (Cystic Fibrosis Trust 2004, Middleton et al. 2014, Moran et al. 2018). There is no evidence to support the benefits of one insulin over another and choice is assessed on an individual basis taking into account lifestyle and clinical and nutritional status (Cystic Fibrosis Trust 2016, Bridges et al. 2018). Insulin has additional anabolic properties and it offers wider flexibility due to the variety of preparations available. In clinical practice within the author's CF centre insulin analogues (aspart, lispro) are primarily used to treat post-prandial hyperglycaemia, they may be taken on their own or as part of a basal-bolus regimen. Basal insulin (detemir, glargine) may also be used on its own to treat insulin deficiency in the early stages of CFD. Insulin therapy is associated with improvements in glycaemic control, lung function and nutritional status (Moran et al. 2009, Hameed et al. 2012, Moheet & Moran 2017).

#### 1.8.6.2 Oral antidiabetic drugs

Oral antidiabetic drugs are not widely used in the treatment of CFD and studies on their use are limited (Ballmann et al. 2014, Onady & Stolfi 2016). A recent study concluded that Repaglinide, a prandial blood glucose regulator, should be considered as an alternative treatment to insulin for patients with newly diagnosed CFD (Ballmann et al. 2018). This was a multi-centre, open-labelled, randomised trial involving 49 CF centres in Europe conducted over nine years. There was no control group to compare the effects of insulin with Repaglinide, so it's uncertain if the small changes observed were due to the effects of the treatment or the natural course of the cohort. A short-acting insulin was used as the comparative, not an insulin analogue, which is most commonly used in routine clinical practice; therefore, not an ideal comparator. The doses of insulin used were also low and this may not have therefore resulted in any significant change. More research is therefore needed before the wider use of Repaglinide is adopted.

Incretin modifiers, such as dipeptidyl peptidase-4 (DPP-4) inhibitors are used in the management of type 2 diabetes. DPP-4 is an enzyme which destroys the hormone incretin. Incretins help the body stimulate the release of insulin when it is required (after meals) and decreases the manufacture of glucagon by the liver when not required (during digestion) (Diabetes UK 2018). Therefore, DPP-4 inhibitors help regulate blood glucose by protecting incretins from damage. DPP-4 inhibitors only work if the pancreas has some functioning β cells (Pathak & Bridgeman 2010). There is limited experience of the use of these products in people with CF (Hellman et al. 2014, Sunsoa et al. 2017, Farrell et al. 2018). These small observational studies have demonstrated that DPP-4 inhibitors are safe, well tolerated and improve glycaemic control. DPP-4 inhibitors may be beneficial for those with CF who have a good nutritional status, early stages of dysglycaemia and demonstrate barriers to insulin therapy (Sunsoa et al. 2017, Farrell et al. 2018). DPP-4 inhibitors are a lower treatment burden than insulin because they are once daily oral agents and the risks of hypoglycaemia are low so routine blood glucose monitoring is reduced.

With the ageing CF population and the improved nutritional status, including the emergence of overweight and obese people with CF, it may be time to start challenging if insulin is the most appropriate treatment for all people with CFD. Oral antidiabetic agents may be a more suitable choice for overweight and obese adults with glucose dysregulation. However more research within this area is required to provide evidence to support this.

#### 1.8.6.3 Nutritional management

Conventional healthy dietary recommendations for the treatment of diabetes are not always appropriate for the management of CFD; diet alone is rarely used to treat CFD. Through the combination of CGMS or serial blood glucose monitoring and a food intake diary the cause of glucose excursions can often be identified. Where glucose excursions are attributed to specific dietary factors such as refined carbohydrate drinks and snacks, which offer very little nutritional benefit, individuals will be advised to either omit them from their diet or change the time they are taken. These dietary

changes have resulted in improved glycaemic control without the need for medication (Barrett et al. 2017). Dietary advice is therefore individualised but based around regular meals and snacks containing complex carbohydrates with modification to the quantity, distribution and timing of simple carbohydrates.

The role of carbohydrate counting, where insulin doses are matched to the carbohydrate content of foods and drinks, is not formalised in the management of CFD. Some individuals may be taught it along with insulin dose adjustment, but the decision is usually based on individualised assessment. Carbohydrate counting and insulin dose adjustment has been shown to result in more dietary freedom in people with type 1 diabetes (DAFNE Study Group 2002), such work does not currently exist, to the same extent, in the CFD population with only a few conference abstracts exploring carbohydrate counting (Robb et al. 2009, Watson et al. 2012, Still & Philip 2017).

One of the challenges to the management of CFD is in those individuals who require oral nutritional supplements or enteral tube feeding to meet their nutritional requirements (Wilson et al. 2000). Both oral nutritional supplements and enteral tube feeds are high in carbohydrates and will therefore increase blood glucose levels. Individuals are encouraged to take oral nutritional supplements as part of their meals, with additional insulin factored in. The introduction of enteral tube feeding requires regular blood glucose monitoring and adjustments to insulin (type and/or dose) to achieve optimal glycaemic control (Kelly & Moran 2013). Through clinical experience patients who require enteral tube feeding tend to be the sicker ones who spend more time in hospital experiencing recurrent respiratory exacerbations. This concurrent instability in health further adds to the complexities in trying to optimise glycaemic control.

People with CFD should be treated at specialist CF centres which have expertise in both CF and CFD, and receive individualised dietary advice and appropriate medical treatment (Cystic Fibrosis Trust 2004, Middleton et al. 2014, Bridges et al. 2018, Moran et al. 2018).

### 1.8.7 Psychosocial

The diagnosis and onset of diabetes in people with CF signifies the development of a second chronic illness, which comes with its own burden of treatment and monitoring in addition to the high treatment burden already associated with managing CF. Only one study was identified which explored health related quality of life specifically in people with CFD. Worse treatment burden domain scores on the Cystic Fibrosis Questionnaire -Revised (CFQ-R) were seen in people with CFD on insulin when compared to those with: CFD not on insulin, impaired glucose tolerance or normal glucose tolerance (Kwong et al. 2019). The score was adjusted for other medication including pancreatic enzymes, nebulised and oral medications. Thus, indicating a significantly greater treatment burden in people with CFD on insulin. It however did not explore the specific elements of the demands associated with increased treatment burden such as blood glucose monitoring, the type and mode of delivery of insulin or the impact of additional clinic attendance. These factors also need to be considered. A generic CF quality of life questionnaire such as the CFQ-R is not designed to identify CFD specific factors, a CFD specific quality of life measure is required. Currently such a patient reported outcome measure does not exist.

A significant part of managing diabetes involves self-management; people with CFD must make decisions about how to manage their life, CF and CFD. In a large international, cross-sectional study, emotional or psychological problems such as anxiety, diabetes distress and depression have been shown to be experienced by at least forty percent of people with diabetes (Peyrot et al. 2005). Diabetes distress is an emotional distress related to the constant anxieties and burdens associated with managing diabetes over time (Fisher et al. 2014). Diabetes distress is associated with sub-optimal glycaemic control and self-management problems (Fisher et al. 2013, Hessler et al. 2014). Very limited research exploring how individuals manage CFD within their daily lives exists (Kelly & Moran 2013), this includes the emotional and psychological well-being of people with CFD.

NICE has published recommendations for the identification, treatment and management of depression in adults with long-term condition which include diabetes

(NICE 2009). However, emotional and psychological support has been identified as one of the '15 healthcare essentials' for people with diabetes indicating that emotional distress should be addressed as part of routine care for all people with diabetes rather than just being treated when identified (Diabetes UK 2016b).

No recommendations exist for the psychosocial and emotional support of people with CFD. NICE CF guidelines recommend people with CF should have access to a clinical psychologist as part of routine CF care and they should conduct a general mental health and wellbeing assessment as part of the CF annual review (NICE 2017). The International Committee on Mental Health in CF recommend the General Anxiety Disorder (GAD-7) and Patient Health Questionnaire (PHQ-9) should be used as screening tools for anxiety and depression in people with CF (Quittner et al. 2016a). The Cystic Fibrosis Questionnaire -Revised (CFQ-R) is the best validated and most frequently used patient-reported outcome measure in CF research (Habib et al. 2015, Ronit et al. 2017). It measures health-related quality of life in adults and adolescents with CF and consist of 44 items across 12 domains (Quittner et al. 2005). The respiratory domain has been approved by the Food and Drug Administration (FDA) in the USA for use as the endpoint in clinical trials (Goss & Quittner 2007). No patient reported outcome measurements specifically for CFD exist.

With earlier diagnosis and treatment of dysglycaemia it is imperative that people with CFD receive sufficient and appropriate support by healthcare professionals, so they can successfully manage their conditions.

## 1.9 Background to diabetes self-management education programmes

As with type 1 and type 2 diabetes a key part of CFD care involves changes in behaviour and lifestyle modifications. For people with CFD this also means the addition of more treatment and monitoring. The focus of care should include the provision of sufficient education and support to facilitate effective self-management.

Diabetes self-management education (DSME) plays a vital role in enabling people with diabetes to manage their illness on a day-to-day basis (Diabetes UK 2015a). DSME programmes should aim to facilitate knowledge, skills and ability in order to improve self-management and clinical outcomes (Beck et al. 2017). People with diabetes need to have the skills, knowledge and confidence to manage their diabetes successfully.

The provision of education of people with diabetes is considered on three levels (Diabetes UK 2015a):

1. One-to-one advice and information
2. Informal ongoing learning e.g. peer groups
3. Structured education programmes meeting agreed criteria (NICE 2016)

NICE (2015) suggest that structured education should form an integral part of type 1 diabetes care and that all adults with type 1 diabetes should be offered a place on a DSME programme. DSME programmes should: be evidence-based, have specified aims and objectives, follow a structured written curriculum with theoretical underpinning, support the development of self-management of diabetes, be delivered by trained facilitators, be quality assured and audited regularly (NICE 2016). DSME programmes are regarded as complex interventions because they contain many interacting components with variable levels of complexity (Craig et al. 2008).

In the UK, there are a wide variety of DSME programmes for people with type 1 and type 2 diabetes (Diabetes UK 2015b). There is not a 'gold standard' DSME programme, but incorporating psychosocial and behavioral strategies into programmes have shown improved outcomes (Funnell et al. 2012). DSME programmes differ in content, theory, format, presentation (individual, group or on-line) and duration. Some programmes are specific to individual hospital trusts and only available to patients who attend that trust while others are available nationally. Programmes which meet NICE criteria, available at a national level, include: Dose Adjustment For Normal Eating (DAFNE Study Group 2002), Diabetes Education and Self-management for On-going and Newly Diagnosed -DESMOND (Davies et al. 2008), X-PERT (Deakin et al. 2006), Diabetes Manual (Sturt et al. 2006a, Sturt et al. 2008, Huxley et al. 2015), Beta Cell Education

Resources for Training in Insulin and Eating- BERTIE (Everett et al. 2003) and Healthy living for People with Diabetes - HeLP Diabetes (Murray et al. 2017)**.** Table 6 highlights the key characteristics of these programmes.

**Table 6 Characteristics of National DSME programmes**

| DMSE | Diabetes type | Format | Delivery | Facilitator |
|---|---|---|---|---|
| **DAFNE**<br><br>(DAFNE Study Group 2002) | Type 1 | Five days delivered over one week or one day a week over five weeks | Group | yes |
| **X-PERT Diabetes, X-PERT insulin**<br><br>(Deakin et al. 2006) | Type 1 or type 2 | Six weekly sessions of approximately 2½ hours | Group | yes |
| **DESMOND**<br><br>(Davies et al. 2008) | Type 2 | One day or two half-days | Group | yes |
| **Diabetes Manual**<br><br>(Sturt et al. 2006b) | Type 2 | 12-week interactive learning programme | 1:1, home | yes |
| **BERTIE (Everett et al. 2003)**<br><br>**BERTIE Online (BERTIE 2019)** | Type 1 | One day per week for four weeks<br><br>Online modular course (not formally evaluated) | Group<br><br>1:1 | yes<br><br>no |
| **HeLP Diabetes**<br><br>(Murray et al. 2017) | Type 2 | web-based self-management programme | 1:1 | Yes |

Both one-to-one and group education has been shown to be effective in improving diabetes self-management (Lawal & Lawal 2016). There is however some contrast between findings and the evidence is limited to a small number of studies. Group education has been shown to be more effective than individual education in improving HbA1c, diabetes knowledge and weight (Deakin et al. 2006, Merakou et al. 2015). In comparison, equal impact on HbA1c at 12-18 months has been seen when comparing group verses individual education (Duke et al. 2009). People often learn from one another in group settings, via observation, copying, and modelling (Bandura 1977a).

Some people benefit from this interactive nature of groups, but this is not applicable to all. NICE (2015) recognise this and advise alternative education programmes of equal standard for people not able or unwilling to take part in a group. What is important is that, however delivered, DSME programmes need to be adaptable to meet specific learning needs of the participants (Lawal & Lawal 2016). For people with CFD DSME programmes will need to be delivered on a one-to-one basis. This is due to strict cross-infection policies that recommend against contact between individuals with CF (UK Cystic Fibrosis Trust Standards of Care Working Group 2011, NICE 2017).

DSME programmes for adults with type 1 diabetes have shown improvements in HbA1c (DAFNE Study Group 2002, Everett et al. 2003), quality of life (DAFNE Study Group 2002), diabetes management skills and food knowledge (Everett et al. 2003). In adults, with type 2 diabetes, reduction in HbA1c (Deakin et al. 2006), greater weight loss and lower prevalence of depression (Davies et al. 2008) have been seen. The availability of DSME for delivery on a 1:1 basis, that have been formally evaluated, is very limited. The Diabetes Manual, a 1:1 intervention for type 2 diabetes, showed small statistically significant improvements in diabetes distress and diabetes management self-efficacy over 26 weeks and reduction in HbA1c maintained at 12 months (Sturt et al. 2006a, Sturt et al. 2006b, Sturt et al. 2008). The first UK trial of a web-based DSME programme for 1:1 delivery to people with type 2 diabetes (HeLP-Diabetes), demonstrated a reduction in HbA1c at 12 months but no overall impact on diabetes distress (Murray et al. 2017). No validated programmes for 1:1 delivery for type 1 diabetes were identified. BERTIE is available as a web-based programme, for people with type 1 diabetes, however it has not been formally evaluated, and its delivery is not structured or facilitated (BERTIE 2019).

DSME plays a vital role in enabling people with diabetes to manage their illness on a day-to-day basis, however no DSME programme that meet NICE criteria currently exists in routine practice for people with CFD. A pilot study of a structured education programme for people with CFD that met NICE criteria has been reported (Watson et al. 2012, Collins et al. 2015). This was based on a modified DAFNE programme and delivered in four sessions, individually, over an eight-week period to 12 participants.

Positive non-significant changes in CF and diabetes quality of life and a trend towards improvement in HbA1c at 6 months were seen (Watson et al. 2012, Collins et al. 2015). However, this programme was very labour intensive for the educators, required a significant amount of participant's time, did not result in many significant sustained outcomes and was difficult to recruit to (unpublished results, S Collins was one of the study educators). Post-programme evaluations (unpublished) were received from nine participants. All found the programme helpful, with learning how to carbohydrate count and adjust insulin to carbohydrate ratios viewed as the most beneficial. Three of the participants suggested that online resources would be helpful. For people with CFD, future DSME programmes need to be more flexible, accessible and adaptable, and more consideration to clinical and nutritional variability and method of programme delivery is required.

## 1.10 My interest in the topic

Prior to commencing this study, I had been working as a specialist dietitian with adults with CF for nearly twenty years. Through my work I developed a strong interest in CFD and its management. I learnt that managing CFD was a complex phenomenon. To support my management skills I developed extended clinical and educational roles such as insulin dose adjustment, carbohydrate counting, setting up CGMS, teaching insulin administration and blood glucose monitoring so that I could work at an advanced level of clinical practice. During this NIHR Clinical Doctoral Research Fellowship I have completed a comprehensive programme of training to develop personal, professional and research skills (appendix 1). This included a supplementary prescribing qualification which allows me, under a clinical management plan, to prescribe insulin for people with CFD.

I learnt that advanced communication and counselling skills were necessary to help manage these patients with multiple co-morbid conditions. I therefore made the decision to complete a MSc in Counselling and Healthcare and Rehabilitation in 2005. My dissertation for this gave me my first exposure to qualitative research methodologies where I explored the impact of the diagnosis of CFD.

The chronic nature of CF and CFD leads to frequent patient contact with healthcare professionals. This helped me start to develop a greater insight into the management of CFD in the context of the lives it affects. I began to appreciate that not everyone with CF has the same relationship with CFD and for some it was a major obstacle and far worse than CF. How CFD impacted upon the lives of people with CFD appeared to affect self-management skills. I became more aware that the requirements of people with CFD need to be considered in the context of how they manage their CF and their lives. This started me questioning why do people experience CFD in different ways and what factors help them or inhibit them in their management and coping?

In the challenging and changing healthcare environment the quality of patient care and support is not always optimal. Time is often very limited and because of this care can often be more reactive rather than proactive. Due to constraints in time and the prioritisation of clinical care needs, educational support often comes second. I found this frustrating as knowledge is the cornerstone for self-management. I began to ask questions about how to improve education of people with CFD without it increasing the time they are required to attend hospital or impacting on their current level of care and support? It also made me think about what are the essential skills needed to be able to effectively self-manage CFD so that time could be focussed upon these?

Through my clinical practice I have worked with lots of different healthcare professionals but their understanding of CFD and its management were very variable. I've witnessed patients being informed of the consequences of sub-optimal glycaemic control with very little or no help being offered on how to improve the situation. I believe that part of this was because some healthcare professionals are not confident enough to give advice because they lack understanding of the complexities of CFD. This led me to question what support and education do people with CFD require and what is the best method to deliver it? It also made me think about the education and support needs of healthcare professionals so they can more effectively help optimise the management of CFD. In the organisation I worked in I felt that many healthcare professionals believed CFD should be managed by specialists in that field. This was of concern to me because CFD it is such a significant co-morbid condition affecting the

lives of approximately half of the adult CF population. These concerns led to a successful application for an NIHR Clinical Doctoral Research Fellowship to undertake this PhD.

My experience in the management of people with CFD has given me the opportunity to present at a national and international levels. I am currently a professional member of the steering group updating the UK CF Trust's guidelines for the management of CFD. I was recently able to influence change through my work, as a consequence of this study. Through sharing my research findings with the steering group they became convinced that the right way forward was to use the term 'cystic fibrosis diabetes' instead of 'cystic fibrosis related diabetes' in our practice. The reasoning behind this is discussed in chapter three but is essentially due to the lack of causal relationship that 'related' implies. The steering group recommended a change in the terminology in their document to CFD.

CFD is associated with increased morbidity and mortality. Insulin is the primary treatment; this requires appropriate training, education and additional self-management activities. DSME programmes have demonstrated effectiveness upon diabetes outcomes in people with type 1 and type 2 diabetes. However, no DSME that meet NICE criteria exist for people with CFD. This study offers the opportunity to better understand the self-management experiences and requirements of people with CFD and identify appropriate methods for the delivery of education and support. This will inform the development of a self-management education programme specific to the needs of people with CFD.

## 1.11 The organisation of the thesis

In this chapter background information about CF, CFD and DSME programmes has been provided. Then an outline of the development of my interests with the management of CFD followed. The remaining chapters are organised as follows:

**Chapter two** provides an overview of the Medical Research Council's (MRC) Framework for developing and evaluating complex intervention and how it has been

used to inform this study. This is followed by a review of methodological design for each stage of the study. The rational for choice of each methodology is explored playing attention to the strengths and limitations of the chosen approaches.

**Chapter three** is a meta-ethnography exploring the experiences of people with CFD in living with and managing CFD. Six studies, a collective representation of 83 adolescents and adults with CFD, were included in this meta-ethnography. A detailed account of the methods used, key findings obtained, and discussion are presented.

**Chapter four** is an empirical qualitative study which uses interpretative phenomenological analysis (IPA) to explore adults with CFD experience of self-managing their diabetes. Eight adults with CFD participated in this study. The method of IPA and key findings are described. Discussion of key findings, which included comparison to current theory and influence upon the MAGIC (managing abnormal glucose in cystic fibrosis) programme development, then follow.

**Chapter five** describes the methods used to develop and review the MAGIC programme. Keys findings from stage one along with the use of a stakeholder development group consisting of people with CFD and healthcare professionals were used to inform the development of the MAGIC programme. Results from the development process give illustrative examples to demonstrate design features and the theoretical grounding of the MAGIC programme. Results from the MAGIC programme reviews are used to demonstrate the face validity of the MAGIC programme. The discussion draws together key findings and the implications for future studies and clinical practice.

**Chapter six** brings together key discussion points from all stages of this study. The contribution of these findings to knowledge and relevance to current literature are considered. The study's limitations, strengths, credibility, implications for practice and further research are then discussed.

Finally, **chapter seven** draws together the conclusions from this study.

# 2 Methodology - the best practice for developing complex interventions

This chapter outlines the methodology for this thesis, the background to complex intervention development and the methodological design for each stage of the study and associated research questions.

## 2.1 Research questions

The overarching research question for this study is “how can we best meet the self-management education requirements of people with CFD?”

Specific research questions for each stage of the study were developed to help answer this overarching research question. These are:

- Stage 1a -what are the experiences of adolescents and adults with CF in living with and managing CFD?
- Stage 1b - what are the self-management experiences of people with CFD?
- Stage 2 – what does a web-based self-management education programme for CFD need to contain?

The overall aim of this study is to develop a self-management education programme for people with CFD.

## 2.2 Complex intervention development

DSME programmes are complex interventions because they contain many interconnected components with variable levels of complexity (Craig et al. 2008). This includes consideration to: content- this needs to specific to type of diabetes, social and cultural needs; method of delivery – is the intervention goup based, 1:1 or web-based; context - is the programme for use in primary care, secondary care or home use; and what level of support does the programme require? The course from development through to implementation of a complex intervention may take a wide variety of different procedures. However interventions for behavioural change need to be developed and evaluated in stages, using an established approach to help ensure they

are feasible, acceptable, safe, effective and efficient (NICE 2007). The failure to demonstrate intervention effect has been seen in many complex intervention studies; this could be a result of true ineffectiveness or unsuccessful intervention design and/or failure to implement the intervention (Levati et al. 2016). The Medical Research Council's (MRC) Framework for developing and evaluating complex interventions is the most widely used guideline (Corry et al. 2013) and was the chosen approach to inform the development of the managing abnormal glucose in cystic fibrosis (MAGIC) programme. The rationale for this is discussed below.

The original version of the MRC Framework (figure 4) consisted of five sequential phases for the development and evaluation of complex interventions (Campbell et al. 2000). Although it was considered hugely influential it was criticised for many reasons, these included: its linear approach, problems with contextual issues (Campbell et al. 2007), and despite highlighting the importance of the early phases of development it lacked details how to achieve stages 1 (review of theory) and 2 (modelling) (Hardeman et al. 2005). A revised version of the MRC framework which aimed to address these limitations was developed (Craig et al. 2008). This is made up of four key phases: development, feasibility/piloting, evaluation and implementation. Figure 5 outlines the main phases and the key functions and actions at each of them, with the arrows representing the main interactions between the phases.

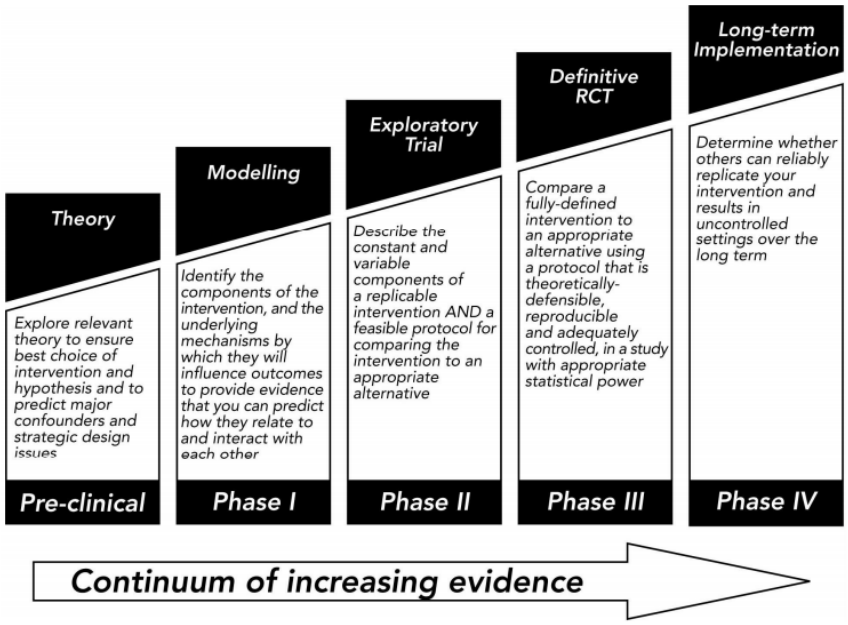


**Figure 4 Original MRC Framework (Campbell et al. 2000)**

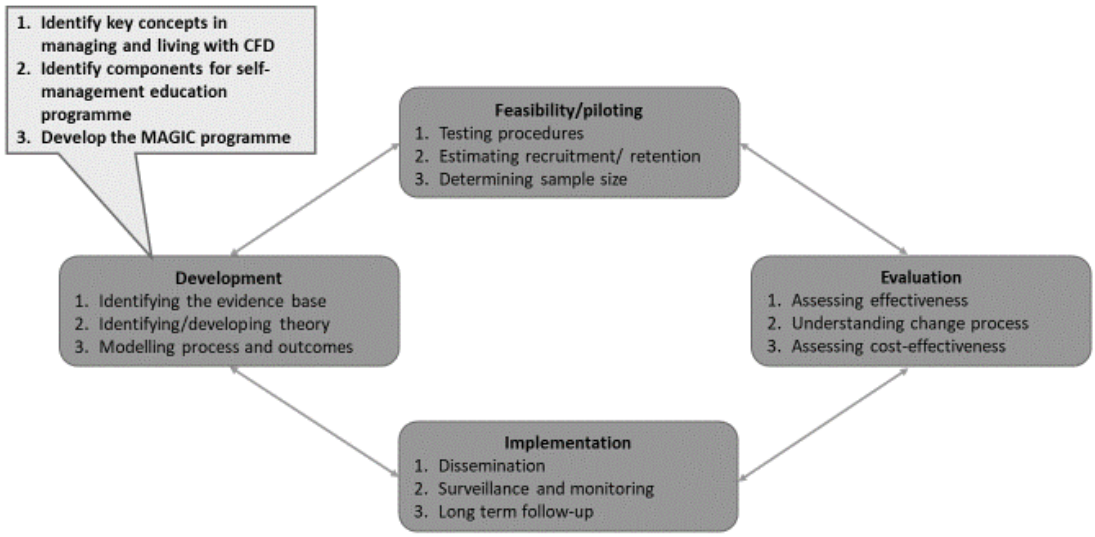


**Figure 5 MRC Framework for developing and evaluating complex interventions (Craig et al. 2008)**

To address issues with the linear approach of the original framework in the revised version there is some overlap between the first three phases as they follow an iterative rather than linear process. The staged approach cannot be removed completely as implementation cannot take place until the first three stages have been completed

and the intervention developed to a point where it will achieve meaningful impact. The issue of context is still not fully addressed in the revised version, with no mention of context in the intervention development stage and only minimal consideration to contextual factors in modifying the effects of the intervention (Moore & Evans 2017). Little consideration is given to examining how interventions interact with context (De Silva et al. 2014). Context is important as it helps shape how interventions work; interventions should therefore not be viewed as distinct packages of different parts described in isolation from their context (Moore & Evans 2017).

The revised version of the MRC Framework expands its description of the many dimensions of complexity, in complex interventions, which include: number of and interactions between different parts within the interventions (control and experimental), number and difficulty of behaviours required by facilitators and participants, number of groups or organisational levels aimed at by the intervention, number and type of outcomes and amount of flexibility permitted of the intervention (Craig et al. 2008).

The development phase of the MRC Framework is the foundation for the intervention. It aims to identify relevant existing evidence base, to identify/generate appropriate theory and model process and outcomes. The feasibility and piloting phase aims to review the acceptability and feasibility of the intervention for which a variety of qualitative and quantitative methods are likely to be required. Refinement of the intervention may be required prior to the evaluation phase which aims to test effectiveness, understand process and assess cost-effectiveness. There are numerous study designs to consider in the evaluation phase these include: experimental designs such as randomised control trials, N of 1 design, or preference trials, however the approach chosen should be guided by the research question and specific characteristics of the study population (Craig et al. 2008). The implementation phase aims to evaluate the long-term implementation and effectiveness of the intervention in the real-world setting. A systematic approach to intervention development and

evaluation through the combined effect of these phases should therefore produce a well-developed, theoretically supported intervention.

### 2.2.1 Rational for choice of MRC Framework

Alternative approaches to the development of health interventions include normalisation process theory (May et al. 2007, Murray et al. 2010) and the process modelling in intervention mapping (Garba & Gadanya 2017).

Normalisation process theory tries to address the gaps that exist between research and its implementation, aiming to identify what is required for successful implementation of interventions into routine practice (normalisation) (May et al. 2007, Murray et al. 2010). There are four components: sense-making (coherence), engagement (cognitive participation), work done to enable the intervention to take place (collective action) and formal and informal evaluation of the benefits and costs of the intervention (reflexive monitoring) (Murray et al. 2010). Normalisation process theory has more focus on context, which is lacking from the MRC Framework, but its main emphasis is on implementational potential and not development, the latter of which is the focus of this thesis. However, it may be worth considering normalisation process theory in the future to help inform the evaluation and implementation of the MAGIC programme.

Intervention mapping is a framework designed to try to bridge the gap between theories and practice. It consists of five stages which follow the course from the identification of need or a problem to the recognition of a solution (Bartholomew et al. 1998). The main focus of intervention mapping appears to be on health promotion/ disease prevention e.g. HIV prevention, breast and cervical cancer screening, influenza vaccination (Garba & Gadanya 2017). These are not multifaceted or considered complex interventions therefore intervention mapping is not applicable to the requirements of the development of the complex intervention for this study.

Regardless of the model researchers should consider three key issues in defining and evaluating complex interventions (Blackwood 2006):

1. Use relevant research evidence systematically in developing the parts of the intervention – use of exploratory research seeking expert views as well as theory and literature
2. Improve the definition and measurement of complex intervention outcomes – use a framework that will make sure relevant outcomes measurements are used to assess all or part of the intervention
3. Adopt relevant research designs required for each part- consider the use of qualitative research methods for clinically related opinions, use of randomised control trials (RCT) to establish cause and effect

In a systematic review aiming to identify the most comprehensive approach to developing complex interventions, in nursing, the only identified guidelines used was the MRC Framework (Corry et al. 2013). With 9 out of 14 papers describing intervention development using this guideline; the remaining five papers did not specify the use of the MRC Framework or name another framework they used to guide their intervention development. Similar findings were seen in a scoping review aiming to identify and synthesise evidence relating to the optimisation of complex interventions at the pre-trial stage where 17 of the 27 studies reported using the MRC Framework (Levati et al. 2016). Lack of regards to contextual issues was a criticism of the MRC Framework (Moore & Evans 2017). Context is an important consideration to in the management of CFD because CFD has to be managed within the context of the competing demands of managing CF and daily life. What is currently unclear is how CF and CFD are managed together and what can be learnt from this to inform the intervention development. There are however no other frameworks for the development and evaluation of complex interventions that provide sufficient guidelines or evidence to support their use. The MRC Framework (Craig et al. 2008) was therefore deemed to be the most appropriate choice to inform this study.

### 2.2.2 MRC Framework applied to the development of the MAGIC programme

To support the design of the the MAGIC programme, this study will focus on the development phase of the MRC Framework, which is concerned with the identification

of the evidence base, developing theory and modelling. There is currently little available evidence, regarding the experiences of living with and managing CFD, to build upon to develop the MAGIC programme. The MAGIC programme will therefore be developed in two stages (figure 6). Qualitative evidence synthesis, qualitative patient interviews and use of people with CFD and healthcare professionals, as part of the MAGIC programme stakeholder development group will provide the evidence for this intervention and guidance for its theoretical underpinning. As a starting point the stages of the MAGIC programme development have been mapped upon the MRC Framework, along with the specific aims and work undertaken at each stage (table 7).

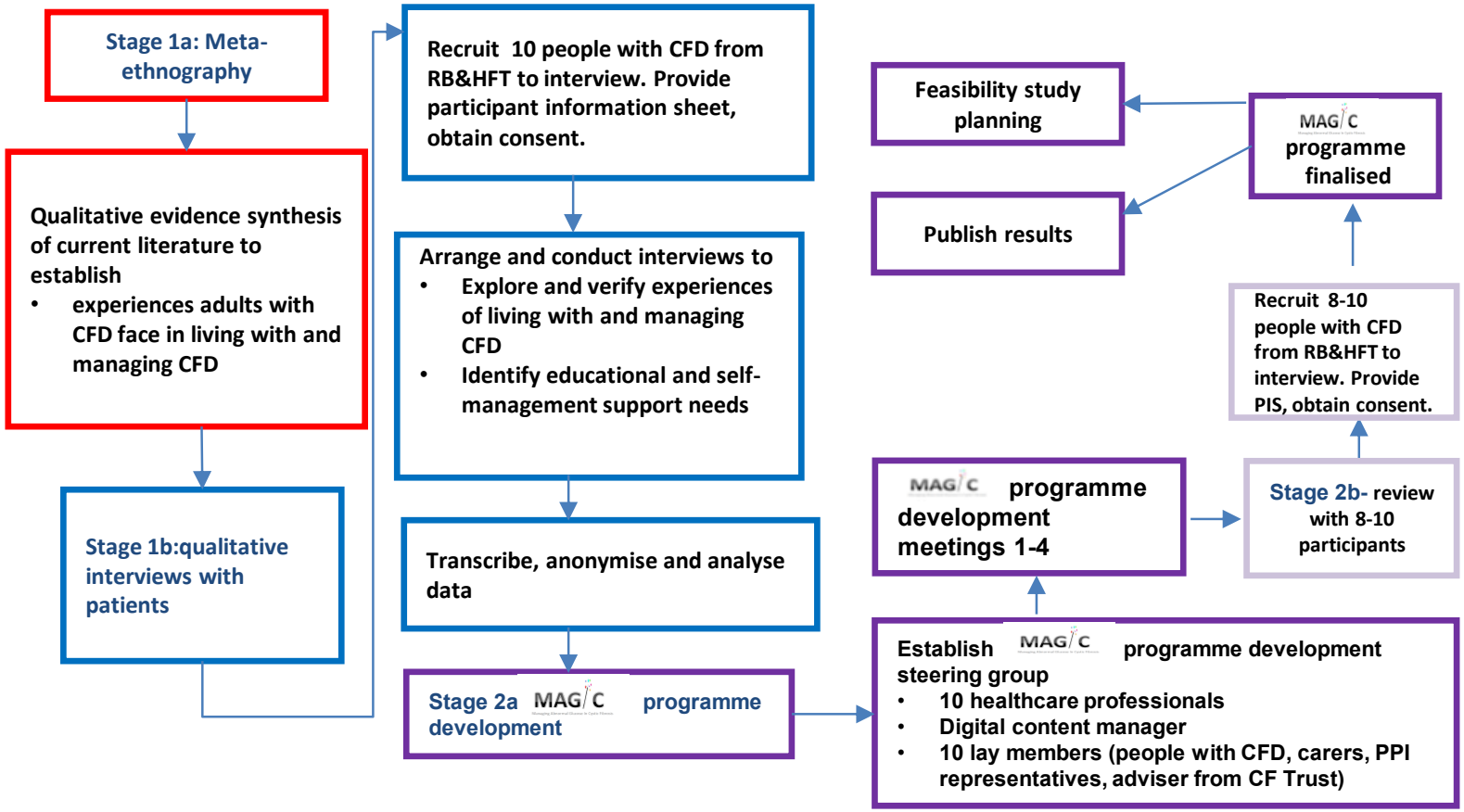


**Figure 6 Study flow chart**

**Table 7 Work undertaken and mapped onto MRC Framework**

| Development phase | | |
|---|---|---|
| | **Aims** | **Work undertaken** |
| **i. Identifying the evidence base** | • To gain a greater understanding of how adolescents and adults with CF live with and manage CFD<br>• To provide a framework to help structure the interview topic guide for the qualitative interviews.<br><br>• To confirm the findings from the meta-ethnography prospectively and to identify any additional concepts<br>• To establish what are the barriers, facilitators and successes that adolescents and adults with CF experience in managing their CFD | i. meta-ethnography of adults' and adolescents' experiences of living with CFD (stage 1a)<br><br>ii. Empirical IPA study with adults who have CFD- identify factors influencing self-management (stage 1b) |
| **ii. Identifying/developing appropriate theory** | • To identify knowledge and skills people with CFD require and use for self-management of CFD and how they might be incorporated into a programme<br>• To help identify content for a CFD self-management education programme | i. Empirical IPA study with adults who have CFD- identify concepts for DSME programme (stage 1b)<br>ii. Literature review to identify theoretical underpinnings of DSME programmes<br>iii. Stakeholder group consisting of lay members and healthcare professionals to co-design the MAGIC programme |

| | | |
|---|---|---|
| **iii. Modelling process and outcomes** | • To assess if the MAGIC programme has face validity (i.e. is a practical, helpful, informative and user-friendly resource) for promoting self-management education of CFD | i. Review of MAGIC programme modules |

## 2.3 Identifying the evidence base using meta-ethnography (stage 1a)

A qualitative evidence synthesis was chosen to answer the question "What are the experiences of adolescents and adults with CF in living with and managing CFD?" with the aims being:

- To gain a greater understanding of how adolescents and adults with CF live with and manage CFD
- To provide a framework to help structure the interview topic guide for the qualitative interviews in stage 1b

A qualitative evidence synthesis was chosen, instead of a narrative literature review, in order to go beyond purely describing and summarising findings. This meta-ethnography will address gaps in evidence by establishing what is known about living with and managing CFD from the current literature. It will develop a deeper understanding of the experiences of living with and managing CFD, which will provide a framework for qualitative patient interviews in the next stage of this study.

### 2.3.1 What is qualitative evidence synthesis?

Qualitative evidence synthesis is the amalgamation of individual qualitative studies to generate a new understanding of the topic being studied. Over the last ten years there has been an increase in the use of qualitative evidence synthesis (Lee et al. 2015). There is, however, no standard method for carrying out the synthesis of qualitative studies. Cochrane Qualitative and Implementation Methods Group have recently published a series of papers to provide guidance on conducting qualitative and mixed-methods evidence synthesis (Cargo et al. 2018, Flemming et al. 2018, Harden et al. 2018, Harris et al. 2018, Noyes et al. 2018a, Noyes et al. 2018b). However more than 30 methods for synthesising qualitative data have been described (Noyes et al. 2018a). Many of these methods are adaptations of four distinct approaches, namely- meta-ethnography, grounded formal theory, cross case analysis and meta-study (Finlayson & Dixon 2008).

Qualitative evidence synthesis methods are generally described as being predominately interpretative or integrative (Campbell et al. 2011). Integrative synthesis produces a description of the subject under study whereas interpretative synthesis produces a new theory of the subject (Paterson 2011). Although the approach to qualitative evidence synthesis can differ in methods, epistemology (theory of knowledge) and terminologies it shares common characteristics such as: involving a research team, arranging the synthesis according to the research objectives and synthesising a number of identified studies (Paterson 2011), where the aim is to identify concepts from the published findings of individual studies and to interpret/integrate them as a collective representation. Noblit and Hare (p28, 1988) describe this as "making a whole into something more than the parts alone imply."

Qualitative evidence synthesis gives an important overview of reported findings around a topic and presents new perceptions of these. Synthesising single qualitative studies together increase the evidence base, to create a more powerful explanation and higher level of understanding (Doyle 2003). Thus, increasing evidence can result in an enhanced transferability of research findings.

The aim and purpose of the qualitative evidence synthesis should inform the appropriate choice of method for synthesis. Researcher's experience, availability of time, audience and type of evidence available also need to be considered when planning a qualitative evidence synthesis (Finlayson & Dixon 2008, Paterson 2011, Toye et al. 2014). These factors were considered when meta-ethnography was chosen as the method of qualitative evidence synthesis for stage 1a.

### 2.3.2 Rationale for choosing meta-ethnography

Meta-ethnography is an interpretative approach to synthesising qualitative studies. It uses the interpretations of data collected through interviews for its synthesis (Noblit & Hare 1988). Unlike raw data for qualitative research such as participant quotes, which are subjected to author's selection bias, all interpretations are published in the research manuscripts. The focus of meta-ethnography on these interpretations means that it remains close to the original study's findings. It is not re-analysis of the raw

data. Through interpretation meta-ethnography tries to make sense of the data rather than being purely descriptive. Thus, achieving a higher level of interpretation and understanding. This is important in trying to understand the experiences of living with CFD where there is a need to understand the experience and also the context of where the experiences occur. The use of meta-ethnography is therefore appropriate to answer the overarching research question of stage 1a, which is - what are the experiences of adolescents and adults with CF in living with and managing CFD? With the aims being to gain a greater understanding of how adolescents and adults with CF live with and manage CFD and provide a framework to help structure the interview topic guide for the qualitative interviews.

Meta-ethnography is one of the most established and referenced method of qualitative evidence synthesis (Campbell et al. 2011). The original focus of meta-ethnography was with only ethnographic studies however, it is now used to synthesise many qualitative research methodologies (Atkins et al. 2008). It has been widely used across healthcare research, social sciences and medicine (Campbell et al. 2011, Prorok et al. 2013, Woodman et al. 2014).

The core of meta-ethnography is translation (Britten & Pope 2011). This is the analysis process which systematically compares and contrast the concepts from different qualitative studies. Noblit and Hare (1988) p38 describe translation as “one case is like another except that …” Three types of translations are described: reciprocal (similar), refutational (contradict) and line of argument (what can be said about the whole based on the parts) (Noblit & Hare 1988). Through synthesising translations, a higher-level interpretation of all the primary studies is achieved. Thus, a systematic and in-depth understanding of how the primary studies are related is achieved through the comparison of concepts within and across the studies.

### 2.3.3 Criticisms of meta-ethnography

Meta-ethnography has received several criticisms (Atkins et al. 2008, Toye et al. 2014, Lee et al. 2015). First limited guidance has been given on search strategies (Dixon-Woods et al. 2005, Toye et al. 2014). In their original work Noblit and Hare (1988) did

not advocate an exhaustive search strategy. They believed the conceptual insight of meta-ethnography is the focus and incorporating large number of studies removes the focus from this (Noblit & Hare 1988).

Secondly what studies should be included. Noblit and Hare's meta-ethnography (1988) pre-dated quality appraisal. However, they recommended the inclusion of studies judged upon the usefulness of the findings. There is now a trend towards appraising the quality of studies to be included in a qualitative evidence synthesis, using tools and/or checklists from organisations such as the Critical Appraisal Skills Programme (CASP) (Britten et al. 2002, Pound et al. 2005, Atkins et al. 2008, Toye et al. 2014). There is however no consensus which tool should be used (Campbell et al. 2011). The CASP criteria for quality appraisal is widely used within the NHS (CASP 2016), is easily accessible and each criteria is clearly defined. This is particularly helpful to a novice qualitative researcher, like myself, who benefitted from the clear guidelines. Modified versions of CASP have been applied to meta-ethnography studies (Campbell et al. 2003, Malpass et al. 2009). These contain additional screening questions which ask: does the study report results from qualitative research and did it involve both qualitative methods of data collection and analysis? and is this research applicable to the synthesis topic (Pound et al. 2005). Answering no to either of these pre-cursor questions would lead to exclusion. These screening questions would be particularly beneficial in qualitative evidence synthesis involving large numbers of studies as they quickly rule out unsuitable studies. One of the major disadvantages of quality appraisal tools is their focus on research methodology and design. Thus, if exclusions are based on this, concept rich studies may be missed (Atkins et al. 2008, Toye et al. 2014). The modified CASP was chosen to appraise the methodological quality of the studies included in this meta-ethnography because of its accessibility, ease of use and previous use in similar meta-ethnographies (Campbell et al. 2003). However close attention would be paid to the richness and depth of data in the qualitative studies, particularly the meaning it has for people with CFD, and studies would not be excluded on the basis of methodological flaws alone.

The use of meta-ethnography was deemed appropriate to meet the aims of this qualitative evidence synthesis because of its focus on understanding and meaning and its established use as a form of qualitative evidence synthesis in health and illness research. This synthesis was therefore undertaken using Noblit and Hare's (1988) original meta-ethnography method. Further considerations were given to more recent interpretations of meta-ethnography, which included: is qualitative synthesis suitable, is a synthesis needed, is there an appropriate team of researchers, identifying the search strategy, using a systematic search and the role of quality appraisal (Atkins et al. 2008, Toye et al. 2014). Details of specific methods used will be presented in chapter three.

## 2.4 Identifying the evidence base and developing theory using IPA (Stage 1b)

Research methodology tends to be classified as either quantitative or qualitative. Qualitative research is exploratory and inductive; knowledge is acquired from those experiencing the phenomena in their social world. Whereas, quantitative work is concerned with cause and effect and hypothesis testing (Finlay 2011, Lyons & Coyle 2016). A quantitative research design such as a national survey of the experiences of people with CFD would not provide a sense of meaning of the experience for the individual. It would generate more numeric and categorised data which would be deficient in personal meaning. A qualitative approach will capture the depth, and not just the breadth, of the experiences. This means not just trying to understand the experiences of self-managing CFD, but also the context and circumstances in which they occur. A qualitative approach was therefore deemed the most appropriate to answer the question "what are the self-management experiences of people with CFD?" Finlay (2011, p8) states:

> "Qualitative research illuminates the less tangible meanings and intricacies of our social world. Applied to the therapy field it offers the possibility of hearing the perceptions and experience of service users."

This study will build upon the findings from the meta-ethnography and seek to gain a greater understanding of the barriers, facilitators and requirements for the self-

management of CFD. This will lead to a better understanding of what self-management experience of CFD means to participants, from their perspective.

### 2.4.1 Philosophical underpinning of qualitative research

Mills & Birks (2014, p 18) describe philosophy as "a view of the world encompassing the questions and mechanisms for finding answers that inform that view." An individual's philosophical beliefs will therefore guide the approach they take to decipher problems in their daily life. How the social world is studied raises a number of philosophical questions, particularly with regards to epistemology (study of knowledge) and ontology (nature of reality).

Ontological assumptions are concerned with what is reality; these are the perceptions of how things really are and how things really work, thus contributing to the understanding of existence (Lyons & Coyle 2016). How will the interplay between CF and the diabetes be lived by study participants and how many different realities of this interplay will emerge? Ontological assumptions range along a continuum from realism to relativism (figure 7). Realists argue there are elements of our world that exist in their own right, separate from people's beliefs and understandings of it (Ritchie et al. 2013). In this paradigm, CF must be most important, compared to CFD, because maintaining respiratory health in a major factor in survival for people with CF and must be the priority of self-care. Thus, there is a difference in the way the world is and the meaning and interpretation of that world viewed by the individual; phenomena have an independent existence which can be discovered via research. Relativists argue that reality does not exist independently of people's beliefs and understandings; it is reliant on the ways we come to know it (Ritchie et al. 2013). Reality is individually created through the interactions between language and aspects of the world (Lyons & Coyle 2016). Thus, meaning is constructed in and through language and language used. People with CFD will have had CF for many of years prior to its diagnosis. They will be developed experiences in healthcare systems and in managing treatment regimens, this will influence subsequent experiences and management decisions.

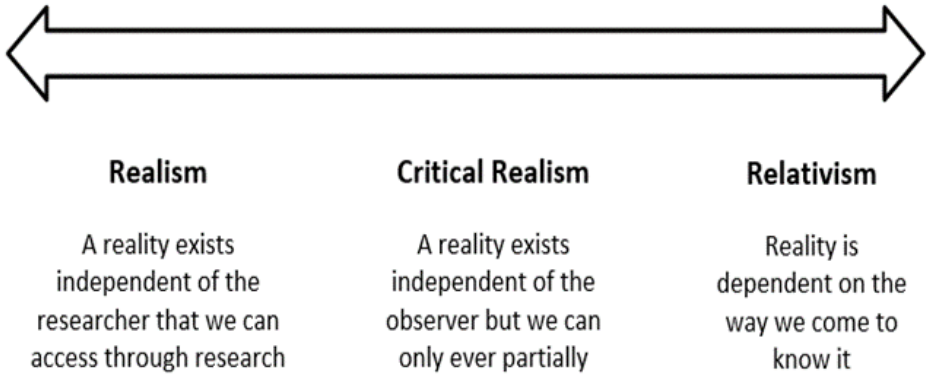


**Figure 7 Ontological continuum (Braun & Clarke 2013)**

Epistemology is the branch of philosophy concerning the theory of knowledge; it attempts to answer the question how and what, can we know? (Lyons & Coyle 2016). Guba and Lincoln (1994, p108) suggest epistemology questions "what is the nature of the relationship between the would-be-knower and what can be known?" Epistemological views include: objectivism, subjectivism, dualist and constructionism (Guba & Lincoln 1994). Epistemology is frequently considered in conjunction with ontology.

Research paradigms are the frameworks that represent the shared perspectives of how the world is viewed and what knowledge is generated from that perspective; they guide research conduct and decision making (Silverman 2013). Many research paradigms have been described, they include: positivism, post-positivism, post-modernism, critical theory and interpretivism/constructivism (Mills & Birks 2014). Research paradigms are characterised according to their ontology, epistemology, methodology (how to acquire knowledge and conduct research) and methods (Guba & Lincoln 1994). Thus, generating a holistic representation of how knowledge is viewed, how researchers view themselves in relation to this knowledge and the methodological approaches used to discover it.

Quantitative research frequently takes place in a positivist paradigm, which adopts an ontological position of realism - where objects/ events/ phenomena have an existence, independent of the researcher, which can be discovered via research (Scotland 2012). Epistemology is objectivism where the goal of research is to produce an understanding

of an objective reality which is impartial and unbiased (Lyons & Coyle 2016). Research methodologies are scientific; aiming to establish cause and effect (Creswell 2008) but lacking in personal meaning and understanding.

Although qualitative research takes place within many research paradigms the interpretivist paradigm is the major contributor. The interpretivist paradigm adopts an ontological position of relativism, where reality is viewed as subjective, it is individually constructed and thus varies from person to person (Guba & Lincoln 1994, Creswell 2008). Epistemology is subjectivism where meaning is created through the interaction between consciousness and the world (Scotland 2012). Research methodologies are interpretative aiming to understand the phenomenon of interest from an individual's perspective, exploring an individual within the social world they live in. People with CF and CFD are very a diverse population in terms of clinical status, age and social background and thus will have variable experiences constructed from the world they live in.

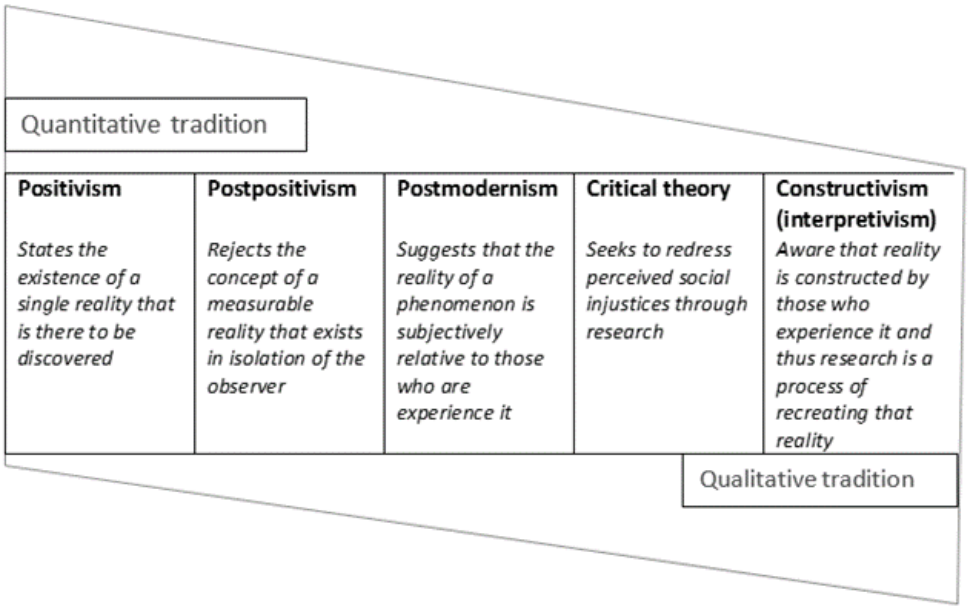


**Figure 8 Philosophical assumptions of research paradigms (modified from Mills & Birks, 2014)**

The relationship between philosophical assumptions and research paradigms is summarised in figure 8, this is not fixed in nature and can be considered as a continuum (Mills & Birks 2014). There is a move from the objective epistemology and

realist ontology of an ordered and structured world associated with positivism towards the more subjective epistemology and an ontological view of a world of multiple realities constructed by humans associated with interpretivism. Critical realism takes a position mid-way between positivism and constructivism/interpretivism, adopting a perspective that while a reality exists independent of the observer, the reality cannot be known with certainty (Mills & Birks 2014). Critical realists holds a realist ontological perspective (belief a real world exists independently of our beliefs and constructions) with a constructivist epistemology (our own knowledge and understanding of this world is inevitably a construction from our own point of views) (Maxwell 2012). Critical realist try to generate knowledge, through interpretation, that captures and reflects as truthfully as feasible a phenomenon that is happening in the real world (Willig 2013). It is acknowledged that multiple and subjective realities and understandings of the world exist instead of one single objective reality (Holloway 2008). Critical realists view individual's beliefs and meanings as equally real to physical objects and processes thus, believing the meanings people attach to things have influences on their actions and for their physical world (Maxwell 2012). Thus, critical realism assumes our ability to know about reality is not perfect and therefore we aim to achieve the best understanding possible.

The approach of critical realism has been adopted within stage 1b. Its location is within the interpretivist framework where the importance is in the understanding of individual's perspectives in managing their illness within the context with which they live their lives. Trying to understand the experiences of managing CFD from the individual's perspective is really important to allow for the diversity seen self-management experiences of people living with CF and diabetes. This information will inform the development of a self-management education programme, so it is important to understand what variability exists so it can be addressed in the programme.

In critical realism there is the ontological view that reality is seen as something that exists independently of those who observe it but, it is only available through the insights and interpretations of the individuals experiencing it. The significance of the

insights and interpretations of the participants in the generation of varying viewpoints and understanding is acknowledged with the aim to discover the complexity and depth of reality of their experiences. While acknowledging that interpreted knowledge is not totally objective it reflects some truth about the phenomenon, but it is influenced by our own interpretations. Figure 9 represents a philosophical overview of stage 1b.

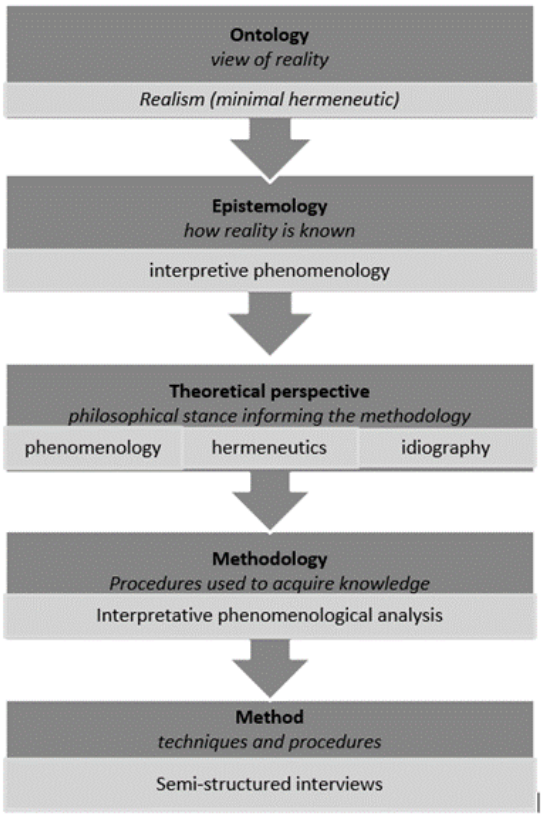


**Figure 9 Overview of philosophical foundations for this study**

## 2.4.2 Interpretative Phenomenological Analysis

A qualitative research design using IPA was chosen to answer the question "what are the self-management experiences of people with CFD?" With the aims being:

- To confirm the findings from the meta-ethnography prospectively and to identify any additional concepts
- To establish what are the barriers, facilitators and successes that people with CF experience in managing their CFD

- To identify knowledge and skills people with CFD require and use for self-management of CFD and how they might be incorporated into a programme
- To help identify content for a CFD self-management education programme

In 1996 Jonathan Smith introduced IPA as a qualitative methodology in health psychology as an alternative to other established qualitative methodologies such as grounded theory (Smith 1996). IPA is concerned with exploring and understanding, in depth, the personal lived experience and how individuals make sense of this (Smith et al. 2009, Shinebourne 2011). Thus, IPA attempts to understand what the world is like from the participant's perspective. Most IPA work tends to be health, illness or impairment focussed (Smith et al. 2009).

#### 2.4.2.1 Theoretical underpinnings of IPA

Smith 2009, p46 states "IPA is not trying to operationalise a specific philosophical idea, but rather draws widely, but selectively from a range of ideas in philosophy." IPA is based on three key areas of the philosophy of knowledge: phenomenology, hermeneutics and idiography.

##### *2.4.2.1.1 Phenomenology*

Phenomenology is a philosophical approach to researching experience, particularly the meaning of experiences for individuals (Smith et al 2009). IPA is significantly influenced by Heidegger's phenomenology work 'Being and Time' which represents a phenomenological perspective where our world around us and our experiences are joined. Thus, a person's place in the world is always in relation to something, temporal and influenced by perception. Larkin et al (2006, p107) describes Heidegger's position acknowledging "that nothing (literally no-thing) is ever revealed as anything (real or unreal, thing or no-thing) except when we encounter it and, hence, when it is brought meaningfully into the context of human life." Therefore, any findings made are inevitably a function of the relationship that is created between the researcher and topic/object of study.

IPA acknowledges that experience cannot be accessed directly from individual accounts and therefore the interpretation of people's meaning-making activities is

essential to help understand the phenomena (Smith 2009). For people with CFD this would include decisions about insulin doses, food choices, checking blood glucose levels and making treatment decisions such as omissions or prioritisation. Through the use of unstructured interviews and using open-ended questions such as “can you describe how you learnt to manage your CFD?” participants will be given the opportunity to speak in depth, in their own words about their experience of self-managing CFD thus allowing their ‘being and time’ to be uncovered. The perspectives on meaning unique to an individual’s relationships through being embodied and situated to the world begin to unfold. Thus, IPA’s phenomenological element aims to explore how has this phenomenon has been understood by this individual.

#### *2.4.2.1.2 Hermeneutics*

Hermeneutics ‘the theory of interpretation’ is the second major theoretical underpinning of IPA (Smith et al. 2009). Schleiermacher, a hermeneutic theorist, influence on IPA highlighted the significance of context. Interpretation involved both grammatical (exact and objective textual meaning) and psychological (author’s unique intentions) elements (Smith et al. 2009). Thus, meaning is available for interpretation but the wider context in which the text (transcript) was produced needs to be considered (Smith et al. 2009). An understanding of the participant as well as the text and a perspective upon the participant’s meaning of the text is trying to be achieved. Smith et al (2009, p23) claims its relevance to IPA is that it “might offer meaningful insights which exceed and subsume the explicit claims of our participants.” Thus, through the interpretative process a perspective on the text, which the author does not or cannot offer, is achieved. For a population of people with CFD, with many diverse characteristics, it is important to consider their unique context within which they exist and the social, emotional, physical factors that influence it, such as the additional treatment and emotional demands required from someone with CF to be able to effectively manage their CFD whilst unwell with a respiratory infection. Continually questioning oneself when analysing the text involves interpreting what the individual is trying to say beyond describing the experience and attempts to interpret in relation to social, cultural and theoretical constructs.

Heidegger's work was again a major influence (Larkin 2006). He acknowledged that researchers' fore-conceptions (assumptions and ideas) contribute to the interpretative process but suggests priority should be given to new things instead of the researchers' preconceptions (Smith 2009). As a specialist CF dietitian my knowledge and experience is heavily influenced by my 22 years work within this area; the process of IPA thus acknowledges this and its contribution to the interpretative process. Other members of my research team are from different professions and their expertise in behavioural medicine, nursing, respiratory medicine and education will influences interpretative processes. They will challenge my interpretations and provide their own individual contributions. Researchers therefore always brings their fore-conceptions to the interpretative experience; however, these may not be apparent until presented with the new. Smith et al. (2009) suggest this form of bracketing is a cyclical and dynamic process that can only be partly attained. Thus, researchers should remain open to their preconceptions/ assumptions emerging while engaging with the new.

The hermeneutic circle (table 8) is the dynamic relationship between the part and the whole of an experience; to comprehend the part you have to look at the whole and to comprehend the whole you need to look at the parts (Smith et al 2009). Thus, the meaning of a single word only becomes evident when viewed in the context of the whole sentence and the meaning of a sentence is reliant upon the collective meaning of the individual words (Smith 2009). This is a dynamic way of thinking, which is iterative - moving back and forth through the data rather than completely linearly. Thus, double hermeneutics is achieved- the researcher is making sense of the individual making sense of a phenomena through the processes of understanding, clarification and interpretation. The researcher is attempting to interpret what an individual is trying to say going beyond description of the experience.

**Table 8 The hermeneutic circle (Smith et al. 2009)**

| The part | The whole |
|---|---|
| **Single word** | The sentence in which the word is embedded |
| **Single extract** | Complete text |
| **Particular text** | Complete works |
| **Interview** | Research project |
| **Single episode** | Complete life |

Hermeneutics is therefore an important theoretical underpinning of IPA. With significant contributions from Heidegger who viewed phenomenology as an interpretive activity, where IPA is associated with exploring how a phenomenon appears, acknowledging the researcher's role in facilitating and making sense of this appearance (Smith 2009). Therefore, IPA's interpretative element aims to understand meaning – what does having CFD mean for this individual, in the context of their life when they are also trying to live with and manage CF?

#### *2.4.2.1.3 Idiography*

Idiography is the third major theoretical underpinning of IPA. It is concerned with the particular and with understanding the meaning of an individual's life instead of trying to make claims within a wider population (Smith 2009). Thus, IPA is concerned with understanding the depth and detail of experiences and recognises that experiences of the same phenomena can be different amongst individuals. IPA creates a theoretical generalisability whereby readers can establish relationships between the findings, their professional and personal experiences and existing literature (Smith and Osborn 2003).

The commitment of IPA to the particular is on two levels:

1. Sense of detail- systematic analysis with depth
2. Understanding how the specific observed phenomena have been understood from the perspective of that particular individual in a particular context

IPA recognises that there is no direct access to pure experience and it therefore aims is to get experience close, where people are seen as 'meaning makers' Smith (2009 p33) states; "the meaning which is bestowed by the participant on experience, as it becomes an experience, can be said to represent the experience itself."

IPA therefore has interpretative (hermeneutic), phenomenological and idiographic underpinnings; where the main aim is to explore how people make sense of their experiences. In summary the key assumptions that guide the process of IPA are (Larkin et al 2011):

- Understanding of the world necessitates an understanding of experience
- IPA researchers produce and explore subjective accounts of other people who are always-already submerged in a cultural, linguistic, relational, and physical world
- To achieve a detailed focus on the particular an idiographic approach is required
- Access to experience is achieved through intersubjective meaning-making
- IPA researchers should recognise and reflect upon their own assumptions and experiences to be able to engage with other people's experiences
- Interpretation is fundamental to IPA; researchers should reflect upon their role in creating these interpretations and ensure they are grounded in participants views

### 2.4.3 Alternative approaches to IPA?

Other qualitative approaches were considered when choosing IPA as the research methodology for this stage. Phenomenology focuses on individual experience and the lived world, IPA shares similarities with hermeneutic phenomenology, but IPA focuses more on the individual's sense-making which is more applicable to the research question for stage 1b which is concerned with individual perspectives and unique meanings. IPA has a more structured set of analytical steps than hermeneutic phenomenology (Finlay 2011), this provides helpful guidance for the researcher in conducting IPA, which is particularly useful to less experienced researchers.

Grounded theory was considered because it's rooted in social science. Grounded theory develops an explanatory level account (influences, beliefs, perceptions, impacts etc e.g. how do people do x or y. The sample size in grounded theory tends to be larger and more heterogenous. Grounded theory is about generalisability and building a theory (model) and focuses more on social phenomenon whereas IPA is more psychological in its approach and focuses on the meaning making. It therefore focuses on action and processes rather than what the experience is like. IPA will be able to help develop a greater understanding of the self-management experiences of people with CFD.

In discourse analysis language is seen as a social performance; the process of constructing reality and not reflecting upon it. Talk is a medium for conveying information and knowledge. It analyses how people use words and language to achieve/do certain things. Both IPA and discourse analysis focus on linguistics but IPA also uses cognitive and affective reactions. IPA therefore also uses linguistics to understand how participants make-sense of their experiences.

Both IPA and thematic analysis focus on themes however, thematic analysis is less detailed and less idiographic. Thematic analysis tends to use larger sample sizes than IPA and focuses on generalisability. Thematic analysis does not stem from any an underlying philosophical theory and can therefore informed by the researchers own philosophical assumptions. Unlike IPA, thematic analysis tends to be more descriptive but this will depend upon the philosophical assumptions adopted. Despite the similarities seen, thematic analysis was not deemed appropriate to address the unique individual meanings IPA would facilitate.

IPA offers the best integrated methodology to explore the ways people with CF make sense of their CFD with their experiences of self-management being the focal point of interest. IPA stays close to meaning and by adopting a smaller sample size it allows for better engagement. IPA facilitates an understanding of 'experience' within its context. As IPA is an interpretative approach the context in which the research takes place is understood when interpreting the data. It is important that CFD is understood within

the complex context in which it is managed, particularly in the context of living with an unstable and unpredictable long-term condition like CF. People with CF manage CFD within their cultural, social and medical prioritisations.

### 2.4.4 Criticisms of IPA

IPA has been criticised for not demonstrating enough depth of interpretation and therefore being more descriptive (Willig 2008, Hefferon & Gil-Rodriguez 2011). The interpretative process in IPA is iterative and dynamic of which the hermeneutic circle is fundamental (Smith et al. 2009). Thus, the researcher needs to aim to go beyond description and through interpretation explore the relationship between the parts and the whole.

The role of language in IPA has also received attention (Willig 2008). Does language convey experience or describe it? Thus, what meaning do the words of an interview transcript tells us more about the ways in which people talk about a specific experience within a specific context rather than about the experience itself? However, IPA acknowledges that pure experience is never accessible, and the aim is to gain insight into experience. 'Meaning making' is achieved through contextual analysis of narratives, metaphors and discourse therefore language is entwined with experience (Smith et al. 2009).

A third criticism of IPA is the suitability of accounts, particularly if participants have the level of articulacy to convey their experiences and the meanings of their experiences rather than just opinions (Willig 2008). IPA recognises a link between dialogue, thoughts and emotional state and realises people struggle to express their thoughts and feelings. It is therefore the role of the researcher to interpret emotional state through what is said and by asking essential questions about what is not (Smith & Osborn 2008). The use of field notes taken during interviews can facilitate this by recording accounts of non-verbal communication such as facial expressions or body language.

Despite these criticisms IPA was considered the most appropriate methodology to explore the self-management experiences of people with CFD. Careful consideration

will be given to the depth of analysis achieved and the meaning behind language used throughout the study.

## 2.5 Developing theory to inform the MAGIC programme development (stage 2a)

From chapter one certain elements about CFD have been establish and need be considered in the development of the MAGIC programme. These are:

- the MAGIC programme should be aimed at adults as the mean age of diagnosis of CFD is 18-24 years
- the MAGIC programme will need to be delivered on a 1:1 basis due to cross infection guidelines advising against contact between people with CF
- insulin is the primary medical treatment of CFD and should therefore be the focus of the MAGIC programme

Stage 2a of the research project will answer the question "what does a web-based self-management education programme for CFD need to contain?" It will aim to develop the MAGIC programme as an e-learning resource, where e-learning is defined as learning carried out via the Internet (Oxford University Press 2019). e-learning was chosen primarily due to the younger age of the CF population, to reduce demands on people's time by limiting additional hospital attendance and because of its suitability for 1:1 delivery (this will be explored in greater detail below).

To inform the theoretical underpinnings and hence methodology for the development of the MAGIC programme four considerations need to be explored. These are: what is self-management, what are the theoretical underpinnings of DSME, how to design self-management interventions and why use eHealth technology?

### 2.5.1 What is self-management?

Self-management encompasses all 'the things people do to help themselves live with a long-term condition' (Self-Management UK 2017). A theoretical framework for self-management of chronic illness describes three sets of tasks that are required: medical management, role management and emotional management (Corbin & Strauss 1988)

figure 10. Medical management includes taking medications, modifying diet and monitoring symptoms. Role management concerns maintaining, changing or creating new behaviors or life roles which are part of adapting to changes in health state for example adjusting to reduced exercise tolerance. Finally, emotional management deals with the emotional consequences of having a chronic illness such as thoughts, feelings and changes in mood which can affect quality of life (Lorig & Holman 2003). Corbin and Strauss' (1988) work is based upon patients' perceptions of their chronic illness experiences- their concerns and problems about their chronic illness. Therefore, self-management education programmes, such as the MAGIC programme, need to address all three self-management tasks and focus on patient perceived problems.


**Figure 10 Self-management tasks**

The main factors influencing the ability of people with type 1 or 2 diabetes to self-manage are (Wilkinson et al. 2014):

- Education- the need for relevant, consistent and comprehensible education for individuals and their significant others to enable self-management
- Communication – both positive and negative experiences that impact upon the ability to self-manage
- Support- healthcare providers and significant others can encourage or discourage an individual in their self-management
- Provider issues – quality of care provision and access to healthcare systems influence ability to self-manage
- Personal factors – individual characteristics such as personal beliefs, culture, physical symptoms, learning experiences, psychological factors and practical issues that impact an individual's ability to self-manage

People living with diabetes face a huge variety of self-management issues on a day-to-day basis. The self-management of diabetes is therefore not static but a dynamic continually evolving process and will vary between individuals.

The most common barrier to self-management in older adolescents and adults with CF is, treatment burden, this relates to the duration, frequency and complexity of treatments (George et al. 2010). It has been reported that people with CF spend approximately 2.5 hours per day conducting CF care (Cystic Fibrosis Trust 2018b) consisting of a median of seven daily therapies (Sawicki et al. 2009). Work and social demands were also considered important and are viewed as conflicts between carrying out CF care and the wish to take part in social events or meet work obligations (George et al. 2010). There is very little research looking at self-management of CFD thus (as discussed above) stage 1b will provide the evidence base for this.

### 2.5.2 What are the theoretical underpinnings of DSME

The MRC Framework highlights the need for a robust theoretical background to provide knowledge into how an intervention works, however it does not specify how to choose the most relevant theory (Craig et al. 2008). Having a strong theoretical background will identify the relevant theoretical constructs to be targeted e.g. empowerment, self-efficacy or well-being. This will also be important when considering what tools are needed to monitor interventional outcomes. A review of evidence of health-related self-management programmes has suggested the importance of tailoring interventions to specific conditions (De Silva 2011). This will thus inform the selection of patients who are likely to benefit from the intervention. As the MRC Framework does not provide information on how to choose the most relevant theory to inform complex intervention development existing DSME programmes were used as a guide.

The focus of DSME is encouraging independence and self-confidence to facilitate effective self-care (Diabetes UK 2015b). Self-management education is different from patient education. The focus of patient education is conveying knowledge whereas self-management education is multifaceted. It is a standardised method to help the

development of many core skills including: problem solving, decision making, identifying resources, taking action and developing partnership with healthcare providers (Lorig & Holman 2003).

As discussed in chapter one there are many DSME programmes that exist within the UK, table 9 lists some of these, that meet NICE (2015) guidelines, and their theoretical underpinnings. There is variation in theoretical underpinnings between the interventions, with no consensus on which theory should be chosen. However, a common feature of DSME programmes are they are person-centred, and they recognise the responsibility for self-management is within the hands of the individual.

**Table 9 UK DSME programmes**

| DMSE | Theoretical underpinnings | Delivery |
|---|---|---|
| **Dose adjustment for normal eating (DAFNE)**<br><br>(DAFNE Study Group 2002) | Therapeutic patient education principles<br><br>(Assal et al. 1985) | Group |
| **BERTIE type 1 diabetes education programme**<br><br>(Everett et al. 2003) | Social learning theory<br><br>(Bandura 1977a) | Groups +/- online |
| **X-PERT diabetes**<br>**X-PERT insulin**<br><br>(Deakin et al. 2006) | Empowerment and discovery learning<br><br>(Anderson 2005) | Group |
| **DESMOND**<br><br>(Davies et al. 2008) | Self-management<br>Empowerment<br>(Skinner & Cradock 2000) | Group |
| **Diabetes Manual**<br><br>(Sturt et al. 2006b) | Self-efficacy theory<br><br>(Bandura 1977b) | 1:1 |
| **HeLP Diabetes**<br><br>(Murray et al. 2017) | Self-management of chronic illness framework<br>(Corbin & Strauss 1988) | Web-based, following initial consultation with facilitator |

Due to the management of CFD requiring insulin the MAGIC programme will share more similarities with type 1, than type 2, DSME programmes. Therefore, the aims and

theoretical underpinnings of these programmes for type 1 diabetes were considered more closely. DAFNE (DAFNE Study Group 2002) and BERTIE (Everett et al. 2003) are both derived from the Düsseldorf intensive insulin therapy programme (Mühlhauser et al. 1987) and share aims of learning how to manage insulin doses compared to carbohydrate intake thus allowing greater dietary freedom. The Düsseldorf intensive insulin therapy programme is guided by therapeutic patient education principles. These recognise that to improve health, patient education involves helping them to develop skills to: enable them to better manage their illness, be able to modify treatments and develop improved coping processes (WHO 1998). However, the quality of therapeutic patient education is dependent upon the abilities and skills of the healthcare professionals who deliver it (Petre et al. 2017). BERTIE (Everett et al. 2003) has been modified to now incorporate social learning theory (Bandura 1977a). This suggests that people learn through observing and copying other people's attitudes, behaviours and the outcomes of these behaviours. Social learning theory is dependent on interactions with others and therefore blends itself better to group learning and would be less suitable to base the MAGIC programme, which will be delivered 1:1.

The X-PERT Insulin programme aims to increase skills, knowledge and understanding of diabetes and helps people make lifestyle choices to manage blood glucose levels more effectively (X-PERT Health 2019). The theoretical underpinnings of the X-PERT programmes are empowerment and discovery learning. Empowerment is a concept to direct patient and healthcare professional interactions. Patients should be well-informed active partners in their care and healthcare professionals should provide patients with appropriate education, support and advice to help them make informed choices to achieve goals and overcome obstacles (Funnell & Anderson 2004). Empowerment theory recognises that someone with diabetes is totally responsible for the self-management of their illness (Funnell & Anderson 2004). Empowerment cannot be given or taught it a process that people do for themselves. Discovery learning sees the learner as the problem solver, who gains knowledge through the use of tools, resources and information (Deakin et al. 2006).

Patient empowerment and therapeutic patient education have an emphasis on the role of a healthcare professional as an educator/ supporter. At this stage of development, it is unclear how much involvement healthcare professionals will have with the delivery of the MAGIC programme; this will be based upon evidence from stage one and views of the stakeholder development group. It would therefore not be prudent to design the MAGIC programme based upon patient empowerment without the evidence to support the role of the healthcare professionals in its delivery.

Reviewing the theoretic underpinning of type 1 DSME programmes did not identify strong evidence to support one theory, over another, to inform the MAGIC programme development. Therefore, patient reported outcomes used to evaluate DSME programmes were reviewed to provide some additional guidance.

X-PERT diabetes (Deakin et al. 2006), The Diabetes Manual (Sturt et al. 2006b), HeLP Diabetes (Murray et al. 2017) and more recent DAFNE trials (Elliott et al. 2012) have all assessed self-efficacy as a patient reported outcome. BERTIE was not formally evaluated as it was modelled upon DAFNE which had undergone extensive validation; instead its impact upon knowledge and diabetes management skills were reviewed (Everett et al. 2003). X-PERT Insulin has also not been formally evaluated, it is based on the X-PERT diabetes programme which was evaluated by randomised control trial (Deakin et al. 2006). Except for the Diabetes Manual (Sturt et al. 2006b) none of the DSME programmes reviewed (table 9) were informed by self-efficacy theory. Thus, demonstrating the expectation that whatever theoretical framework guided the programme development a change is self-efficacy was expected.

The recognition of the role of self-efficacy theory in diabetes self-management behaviours (Mishali et al. 2011) and its wide use as a patient reported outcome measure in clinical trials of DSME programmes influenced the decision to use self-efficacy theory to inform the development of the MAGIC programme.

#### 2.5.2.1 Self-efficacy theory

Self-efficacy is an important factor in influencing diabetes self-management behaviours (Mishali et al. 2011). It is a person's belief in their ability to conduct a

specific activity or behaviour under certain conditions (Bandura 1977b). Self-efficacy is behaviour focussed and plays a significant part in the process of behaviour change (Mishali et al. 2011). There are four main influences on self-efficacy: mastery experience, vicarious experiences, verbal encouragement and psychological state (Bandura 1977b). Therefore, DSME programmes need to address these factors as just giving information or educating will not facilitate successful self-management. Table 10 highlights how these influences may be addressed in self-management education programmes. However, the process of changing behaviour often takes time and support needs to be ongoing. There is evidence to suggest that proactive self-management support and focussing on behaviour change and self-efficacy can improve clinical outcomes (De Silva 2011). The self-efficacy framework is therefore appropriate to help understand and predict self-care behaviours which are relevant to the effective self-management of diabetes (Mohebi et al. 2013).

**Table 10 Influences on self-efficacy**

| Influence on self-efficacy | Activities in self-management education programme |
|---|---|
| **Mastery experience** | Building up skills, knowledge and experience through staged learning<br>Direct experience of successfully mastering a task |
| **Vicarious experiences** | Seeing others overcome a task successfully<br>Mirroring of actions<br>Observations of similar individuals |
| **Verbal encouragement** | Persuasion and encouragement of abilities<br>Constructive feedback<br>Acknowledge difficulties |
| **Psychological state** | Paying attention to and addressing mood, stress, anxiety and any factors affecting the individual |

Improving self-efficacy will enhance an individual's belief in their ability to self-manage their condition. It was believed self-efficacy theory would lend itself better to an e-

learning programme delivered on a one to one basis that provides fewer opportunities for developing relationships with healthcare professionals or others with diabetes.

#### 2.5.2.2 Styles of learning

When considering the theory of self-management interventions, it is also important to consider the population and their learning needs. This intervention will be designed for people with CFD, be delivered via an eHealth platform and meet NICE criteria for DSME (NICE 2016). As it is aimed at adults over 16 years of age, due to this being the most likely age from when a CFD diagnosis will be made, consideration to preferences of learning are important. Not everybody learns in the same way. Four styles of learning have been described these are: visual, aural, read/write and kinaesthetic (Fleming 2001) although not all learners exclusively learn by just one style. Table 11 demonstrates how different learning styles will be addressed in the MAGIC programme. The four different types of learning styles will therefore be considered when developing the MAGIC programme.

**Table 11 Examples of styles of learning in magic programme**

| Learning style | Activities |
|---|---|
| **Visual** | Viewing videos<br>Use of illustrations |
| **Aural** | Asking questions/ facilitator discussion<br>Web-chat |
| **Read/write** | Reading text in website<br>Making notes<br>Completing food, insulin and blood glucose diary<br>Reading instructions |
| **Kinaesthetic** | Completing quizzes<br>Practicing scenarios<br>Carrying out tasks on computer/ tablet<br>Carrying out the task for real |

### 2.5.3 How to design self-management interventions

Self-management programmes are complex interventions aimed at improving the way individuals self-manage their chronic illness to improve their health and well-being (Mills et al. 2014). These programmes are complex interventions because they comprise of different parts, subject areas and formats with which participants will work, over a period of time, to try to incorporate into their daily life (Craig et al. 2008).

The MAGIC programme is for people with CFD, user involvement is therefore a key part of its philosophy to ensure the intervention is user friendly and reflects the real needs of the people it is designed for. Co-design, co-production and co-creating are all terms used to describe working collaborations however they all have slightly different meanings. Co-design is collaborative working to address a problem and identify a solution, whereas co-production attempts to implement the proposed solution and co-creation includes the whole process (McDougall 2012). The MAGIC programme development is therefore a co-creation process but the main focus of this stage of the study is co-design. Feasibility/piloting, evaluation and the full delivery of the MAGIC programme into daily life and thus implementation of the proposed solution is beyond the scope of this thesis.

Other methodologies considered as alternatives to co-design included: participatory action research and experience-based co-design. Participatory action research (PAR) has its primary focus upon working in partnership with communities to reduce health and health inequalities (Baum et al. 2006). Although the focus of the MAGIC programme involves communities (in terms of people with CFD) stage two of the programme was concerning intervention development and face validity, it wasn't looking at change in health. Experience-based co-design brings together patients and healthcare professionals its focus is however on service improvements and not intervention development (The Point of Care Foundation 2019). Despite participatory action research and experience-based co-design both recognising the value of patients and working in partnership they are not applicable to use in the development of interventions.

#### 2.5.3.1 Co-design

Co-design is a form of stakeholder engagement; it combines the opinions, contribution, knowledge and experience of people with different perspectives to address a specific problem (Bradwell & Marr 2008). The process of co-design is based around four components (Bradwell & Marr 2008):

- **Participation**- co-design is a collaboration. A close working relationship is developed between participants who are working together to develop a service/intervention.
- **Development** - co-design is a development process involving the sharing of expertise and knowledge.
- **Ownership and power** – co-design shifts power to the process, there is equality in the contributions from all involved. It is an empowering process due to the shared decision-making across all aspects.
- **Outcome and intent** – co-design process is outcome based.

Best practice for co-design have been described and they include: methodology that supports and encourages all components of the co-design process, creation of an environment that is a safe-space for participations to contribute and communicate and maintaining focus on desired outcome which includes balancing view points and maintaining relevant dialogues (Bradwell & Marr 2008). Thus, these factors need to be considered when embarking on a successful co-design project. A stakeholder development group consisting of people with CFD, healthcare professionals and a digital content manager will be established to co-design the MAGIC programme. To reflect different knowledge and experiences attention needs to be paid to recruiting the healthcare professional representatives from a diverse range of professionals including doctors, nurse specialists, dietitians and psychologists. To reflect different management practices these healthcare professionals will also need to be recruited from different CF centres. Patient representatives will be recruited from within the hospital trust and from other CF centres again to reflect diversity and experiences. The stakeholder development group facilitator will have the important role in maintaining the dynamics of the stakeholder group meetings to ensure all members of the group are actively included so they can share knowledge and experiences and equality within

the group is maintained. An additional challenge for this study will be the use of videoconferencing software in the stakeholder development group meetings to allow all the members with CFD to be present as they are not allowed to meet face to face. The James Lind Alliance top ten research priorities for CF steering group successfully used videoconferencing facilities during its workshops involving people with CF and healthcare professionals (Rowbotham et al. 2018). As a representative on this group I gained first-hand experience of the successful use of videoconferencing in a similar situation and also obtained the idea of appointing a 'link person' to maintain contact with the patient representatives at home.

The main benefits of using a co-design design approach are: the end product reflects customer needs, it generates useful information and it creates a feeling of involvement and ownership (Bradwell & Marr 2008). Incorporating stakeholder participation in the co-designing of complex health intervention will: help explain the health care problem, identify appropriate interventions, ensure relevance and usefulness of the intervention and maximise the acceptability and potential effectiveness of the intervention (O'Brien et al. 2016). Therefore, the end results of co-design will be of value to all stakeholders involved but more specifically to the service users it is aimed at.

### 2.5.4 Why use eHealth technology

Due to the life-limiting effects of CF, the CF population tend to be younger; the median age of people with CF in UK is 20 years (Cystic Fibrosis Trust 2018a). CF is a complex multi-system disease requiring a significant amount of care every day. The chronic nature of CF requires frequent hospital appointments for reviews, with 36% of people with CF reported attending 10 or more hospital appointments in the previous 12 months (Cystic Fibrosis Trust 2018b). However, the CF population is changing with more adults now working or studying. Currently 64.2% of the UK population who are 16 years or older are in work or study (Cystic Fibrosis Trust 2018a). People with CF do not want to take more time off work or study than necessary to attend the hospital especially when it has been shown that an average outpatient appointment (including travel time) for people with CF is 4 hours 46 minutes (Cystic Fibrosis Trust 2018b). People with CF are also recommended against face-to-face contact with others who

have CF to minimise the risk of cross-infection (UK Cystic Fibrosis Trust Standards of Care Working Group 2011, NICE 2017). Considering the age of the population and the demands on care and time the decision was made to develop this intervention using eHealth technology. This format can also provide a vehicle for peer support for a population that are prohibited face-to-face contact.

eHealth is defined as "the use of information and communication technologies (ICT) for health" (WHO 2017). Mobile health (mHealth) is considered a component of eHealth but it specifically relates to use of mobile wireless technologies for public health (WHO 2016). Currently within the UK 96% of adults personally own or use a mobile phone, with 78% of adults using a smartphone (Ofcom 2018). As a nation, a significant number of the population therefore have the accessibility to receive eHealth and mHealth technologies.

mHealth technology increases access to health information, skills and services as well as encouraging positive health behaviour changes (WHO 2016). However, research is limited into what processes and procedures are key to co-design mHealth intervention (Eyles et al. 2016). Key factors to facilitate success of mHealth intervention include: active engagement between healthcare professionals and patients and the incorporation of positive outcomes (Burford et al. 2015). Co-design is therefore vital for development of any mHealth technologies.

A systematic review has shown that information technology-based interventions have been shown to have a positive effect upon glycaemic control and encourage diabetes self-management (Riazi et al. 2015). However, their impact on clinical outcomes such as diet, physical activity, blood pressure and lipid levels are more variable. A different systematic review of internet delivered DSME (Pereira et al. 2015) also demonstrated improvements in glycaemic control. It also found improvements in attendance at clinics but access to online DSME material diminished over time. On a positive note, patients easily adjusted to web-based delivery platforms and the use of asynchronous communication for information such as blood glucose diaries and medication changes worked well. Pereira et al. (2015) suggests that combing communication from

healthcare providers and incorporating peer support within education programmes could help sustain patient interest and engagement in DSME activities and provide a means of social support from peers. This is therefore an important factor to consider in the development of a DSME programme with the role of a facilitator being paramount to help maintain this engagement.

In a systematic review of CF self-management education, improvements in behaviours and knowledge seen were mostly short lived and not maintained over time (Savage et al. 2014). However, none of these interventions were internet based. A theory-based, flexible home learning programme has been shown to be an effective way of providing nutrition education and encouraging behaviour change in adults with CF (Watson et al. 2008) but again this was not delivered via internet technology. The use of information technology in the management of patients with CF has focussed mainly on the use of telehealth. This is the use of technology by patients so that healthcare professionals can remotely monitor data on certain health parameters e.g. oxygen saturations, blood glucose levels (NHS 2013). In CF, telehealth has been used for monitoring symptoms, therapeutic interventions and reviewing adherence to prescribed treatments. However, these studies were mainly small feasibility trials (Cox et al. 2012). One case study of behavioural and nutritional treatment delivered to the family of a toddler via telehealth, demonstrated significant increase in energy intake and expected increase in weight and height velocity (Piazza-Waggoner et al. 2006). There is currently no evidence of self-management education programmes being delivered to people with CF through information technology. Although, digital communication, via Skype, has been used to deliver exercise sessions and clinical reviews to people with CF (Griffiths et al. 2018).

## 2.6 Review of the MAGIC programme - stage 2b

Modelling of complex interventions before conducting a full evaluation is recommended to establish any areas that need refinement or improvement (Craig et al. 2008). Designing the MAGIC programme using a stakeholder group will provide a continual mechanism for feedback and review throughout all stages of development.

The MAGIC programme will also be reviewed by people with CFD towards the end of its development for feedback regarding its face validity and ease of use. Further modelling will be required as part of the next stage of evaluation. This will be a study to determine feasibility of the of the MAGIC programme and to identify which patient reported and clinical outcomes are subject to change.

This stage aims to:

- assess if the MAGIC programme has face validity (i.e. is a practical, helpful, informative and user-friendly resource) for promoting self-management education of CFD

Face validity is a method of assessing, at face value, if a questionnaire, test or resource appears to measure what it claims to; it is assessed through subjective judgement (Keszei et al. 2010). Assessing face validity of the MAGIC programme will therefore determine its appropriateness for people with CFD. Face validity has been assessed through the use of scales (Maribo et al. 2016), questionnaires (Twohig et al. 2017), focus groups (Sturt et al. 2006b) and interviews (Banna et al. 2015). Face validity is determined by the use of scoring systems when using scales/ questionnaires (Maribo et al. 2016, Twohig et al. 2017) or by the application of participant feedback following participant interviews (Sturt et al. 2006b, Banna et al. 2015). As no questionnaires or appropriate scales are available to assess the face validity of a unique intervention like the MAGIC programme participants feedback will be used. Banna et al. (2016) reported face validity was achieved when all the recommendations from the study participants had been addressed. This approach will be adopted when assessing face validity of the MAGIC programme because it will ensure equality by addressing all participant feedback; this will be achievable with the planned participant number and allocated timeframe.

The people with CFD recruited to review the MAGIC programme will be different individuals from those who participated in the MAGIC stakeholder development group meetings and qualitative interviews. This will uncover perceptions and values about the programme from those with no prior personal investment in it. These participants

will review the different modules of the MAGIC programme. Cognitive interviewing techniques will be used to focus on the content of the MAGIC programme modules and the cognitive processes involved in understanding and reacting to them. This method will therefore provide a more detailed account of the participants' understanding which you would not obtain from quantitative methodology such as feedback questionnaires.

Cognitive interviewing has been widely used in testing survey and questionnaire design, initially conducted after the initial design of the questionnaire and before piloting (Collins 2015). However its techniques have been applied in a variety of health research including: the appraisal of web-based applications (Berry et al. 2010), modifying education resources (Anderson et al. 2018), developing patient reported outcomes (Holch et al. 2016), and developing/ evaluating nutrition surveys (Carbone et al. 2002, Fernandes Davies et al. 2016). What these studies share are the significant focus upon comprehension which is a major theory in the cognitive process (Tourangeau et al. 2000). Methodologically there is slight variation in cognitive interview processes with some studies conducting rounds of interviews with different sets of participants and making revisions between each interview (Carbone et al. 2002, Holch et al. 2016, Anderson et al. 2018) whilst others just completed one interview with participants (Berry et al. 2010, Fernandes Davies et al. 2016). There appears to be no consensus when to use which interview approach, but careful consideration needs to be paid to the study aims, participant number, timeframes and size of the resource/ tool/ questionnaire being appraised. The precise format and content of the MAGIC programme will be informed by the stakeholder development group, therefore the decisions regarding the approach to interviews will be made once the programme is developed and ready for review.

The background theory to cognitive processes consist of the following (Tourangeau et al. 2000):

- Comprehension – understanding and interpretation

- Recall – participants search memory for necessary information to answer questions
- Judgement – participants appraise questions and/or gauge response when deciding on an answer
- Response – participants give information in answer to the question

Cognitive interviewing techniques will therefore be used to:

- Establish whether participants understand the content of the MAGIC programme
- Explore if participants can follow information provided
- Explore if participants are willing to act on information provided
- Identify if any information is missing
- Establish how easy or difficult it was for participants to use the MAGIC programme
- Appraise the visual presentation of the website

Therefore, a range of techniques will be used to review participants' experiences of the MAGIC programme modules these include: exposure to test materials, observation, think aloud, task completion and probing (D'Ardenne et al. 2015).

## 2.7 Methodological conclusions

This study aims to develop an evidence-based self-management education programme for people with CFD. This was informed by the MRC Framework for developing and evaluating complex interventions (Craig et al. 2008) along with Blackwood's (2006) three key issues, which are fundamental building blocks in defining and evaluating complex interventions. Relevant research evidence and exploratory research will be used in stage one and 2a to develop theory to inform the MAGIC programme development. Appropriate research design has been used to inform each stage of the MAGIC programme development, with consideration also paid to alternative approaches. Although evaluation is beyond the scope of this study the use of self-efficacy theory will provide a framework for relevant outcome measurement in future studies to assess effectiveness of the MAGIC programme. The MAGIC programme will

therefore be developed with a strong evidence-base, robust theoretical underpinnings and appropriate methodology.

# 3 Identifying the evidence base - Stage 1a- meta-ethnography

## 3.1 Methods

### 3.1.1 Research Design

A qualitative evidence synthesis was chosen to answer the question "What are the experiences of adolescents and adults with CF in living with and managing CFD?" with the aims being:

- To gain a greater understanding of how adolescents and adults with CF live with and manage CFD
- To provide a framework to help structure the interview topic guide for the qualitative interviews in stage 1b

This meta-ethnography was registered with PROSPERO International prospective register of systematic reviews (PROSPERO 2016:CRD42016048524).

Conducting a meta-ethnography involves a series of seven stages that overlap and repeat as the synthesis continues (figure 11). This synthesis was undertaken using Noblit and Hare's (1988) original meta-ethnography method with more recent considerations (Atkins et al. 2008, Toye et al. 2014).

All members of the research team: SC, JS, SW and AJ contributed to the meta-ethnography. SC led the meta-ethnography and conducted the systematic literature review, translation of studies and synthesis of translations. The other research team members contributed to the verification of identified studies and the review and exploration of identified concepts. These roles are acknowledged throughout this chapter with the individual's initials. The key findings were also discussed with the study project management group, which consisted of lay and professional representation.

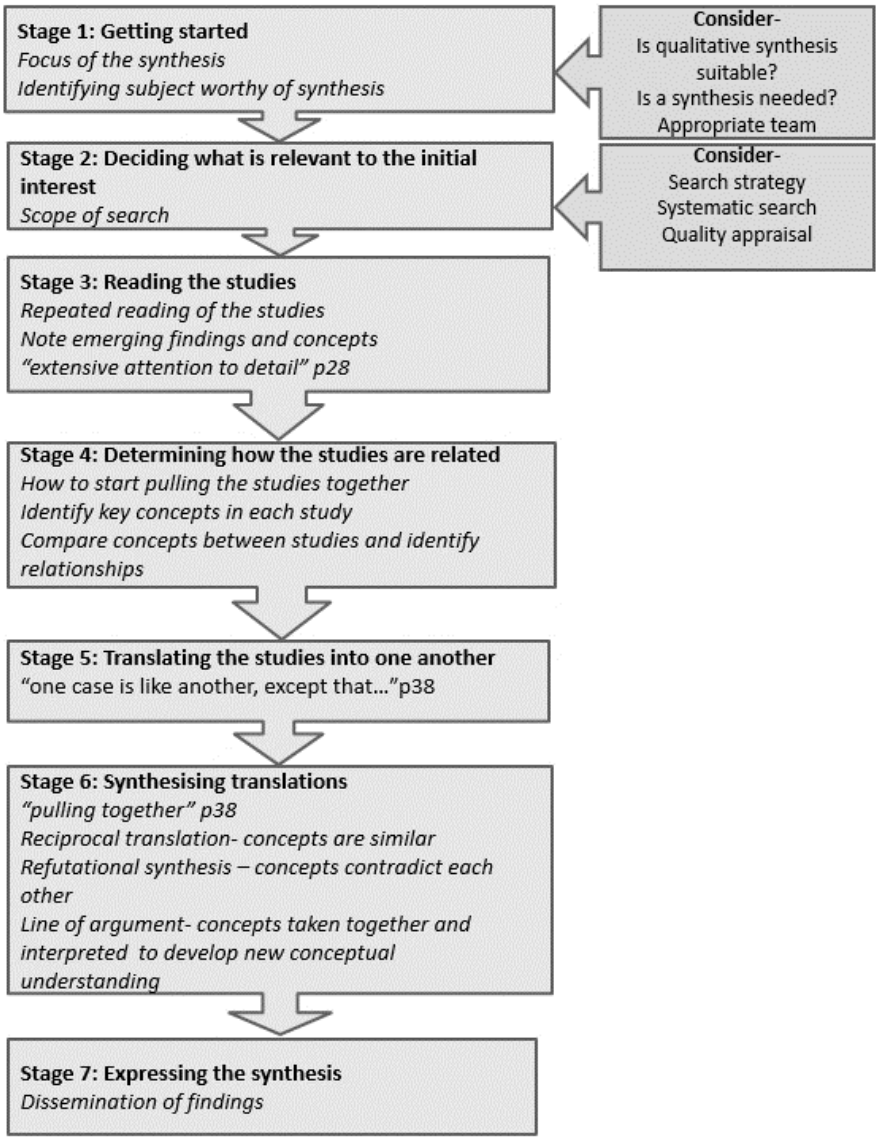


**Figure 11 Seven stages of Noblit and Hare's Meta-ethnography**

### 3.1.2 Stage 1: getting started

Noblit and Hare, p27 (1988) describe this as "finding something that is worthy of the synthesis effort." It also addresses if qualitative synthesis is appropriate, is there the need for the synthesis, what experiences the researchers have and what resources are available (Toye et al. 2014).

This meta-ethnography was driven by experience of working, as a specialist dietitian, with adolescents and adults with CFD; particularly observed difficulties and challenges in disease management, and the significance of worse outcomes for people with CFD.

A qualitative evidence synthesis had not been carried out in this subject area, and was therefore deemed an appropriate subject choice for this meta-ethnography, which was conducted with support from a team of healthcare professionals experienced in qualitative analysis.

### 3.1.3 Stage 2: deciding what is relevant to the initial interest

This involved designing the scope of the search, deciding if an exhaustive literature search is required and then searching for, screening and quality appraising studies. There is a lack of consensus if an exhaustive search strategy is needed for meta-ethnography, however enough data needs to be included so that identified concepts are robust (Toye et al. 2014). Guidelines to how many studies are enough are not available and it is therefore left up to researchers' interpretations.

From experience of working within this specialist area it was known that there was not a wide breadth of qualitative studies exploring the lives of people with CFD, therefore a more extensive systematic literature review was conducted to ensure all evidence was identified.

#### 3.1.3.1 Search Strategy

The sampling involved a systematic literature search of Ovid Medline, CINAHL, EMBASE and PsychINFO, conference abstracts, unpublished thesis and hand searching of references in identified articles to obtain all relevant studies in this area. Databases were searched, by SC, for published journals from 1974 (date data first included in database) to 7th November 2018 (date search updated).

The search strategies (appendices 2-5) aimed to find both published and unpublished studies within this topic area.

1. The initial search was limited to the specialist database Ovid Medline, this is considered the most widely used database in qualitative evidence synthesis (Hannes & Macaitis 2012) and therefore deemed the most appropriate starting point. The search took the following format:

- Free text and subject heading search using keywords cystic fibrosis and diabetes.
- The title and abstracts for relevant papers were screened to identify qualitative studies which addressed the research question “what are the experiences of adolescents and adults with CF in living with and managing CFD? “
- All title and abstracts were screened against the inclusion criteria (SC), this was double checked by JS and SW to ensure all relevant papers were included.
- Where study titles, abstract or key word searches met the inclusion criteria the full text documents were obtained for further exploration.

2. To increase sensitivity and specificity CINAHL, EMBASE and PsychINFO were also searched (SC). These databases contained nursing, allied health professional, health and psychological research papers, which may not have been included in Ovid Medline, thereby allowing access to a more extensive research field. A free text and subject heading search of title and abstracts using key words ‘cystic fibrosis ‘and ‘diabetes’ and ‘qualitative’ or ‘interview’ was conducted. Where study titles, abstract or key word searches met this criterion and possibly addressed the research question “what are the experiences of adolescents and adults with CF in living with and managing CFD?” the full text documents were obtained for further exploration.

3. The reference list of all identified reports and articles were hand searched to identify any additional studies (SC).

4. Conference abstracts proceedings published in Pediatric Pulmonology and The Journal of Cystic Fibrosis, from 2006-2018, were hand searched (SC). These journals publish abstracts from the annual European and North American CF conferences. This timeframe is sufficient to reflect current clinical practice and research. Where any abstracts were deemed relevant to this search the primary authors were contacted for additional information e.g. further publication of results, access to dissertation (SC).

### 3.1.3.2 Inclusion and exclusion criteria

Inclusion criteria:

- Studies with a focus on adults and adolescents, with a mean age greater than 13 years, who have CFD. Greater than 13 years was chosen as this is an age where children are likely to participate in qualitative interviews on their own and start to self-manage their condition
- Qualitative studies concerning the experiences of adolescents or adults living with and managing CFD. This will include the home and hospital environment
- Qualitative findings of mixed-method studies will be considered
- Studies written in English, from any country

Exclusion criteria:

- Studies that focus on the experience of parents/ carers or healthcare professionals
- Studies primarily using quantitative methods
- Studies about type 1 or 2 diabetes
- Studies not written in English

In Microsoft Excel, a worksheet for the search strategy was created. In this all identified studies were recorded along with decisions regarding inclusion or exclusion.

### 3.1.3.3 Quality appraisal

Quality appraisal prior to meta-ethnography synthesis was not advocated (Noblit & Hare 1988). The use of quality appraisal tools in qualitative research synthesis is questioned, as these tools tend to address methodological flaws instead of conceptual strengths (Toye et al. 2014). There is however an increase in the use of quality appraisal tools in qualitative evidence synthesis (Hannes & Macaitis 2012) and therefore a pressure to quality appraise studies. As discussed in chapter two the

modified CASP was chosen to appraise the methodological quality of the studies included in this meta-ethnography because of its accessibility, ease of use and previous use in similar meta-ethnographies (Campbell et al. 2003).

One advantage of using a qualitative critical appraisal tool is that it encourages the systematic and structured reading of studies and therefore helps familiarisation with the data. However, what is needed for meta-ethnography is a tool that facilitates the identification of findings, concepts and interpretations (Campbell et al. 2011). Ensuring transparency by clearly reporting all processes in data collection and analysis is thus important to reflect the completeness of this methodology.

### 3.1.4 Stage 3: Reading the studies

There is a degree of overlap between stages 3-6, and the synthesis involved going back and forth between each stage, exploring ideas and looking for patterns across data sets that were applicable to the focus of this synthesis.

Stage 3 entailed reading and re-reading the identified studies, in detail, to become familiar with the content and to start the process of establishing emerging concepts. It was important to consider what data should be extracted and how should it be done (Toye et al. 2014). Noblit and Hare (1988) use the terms metaphors/ concepts, phrases or ideas however, it can be hard to distinguish what is meant by these terms. It is suggested that concepts are more meaningful ideas, which have some analytical properties, whereas findings are more descriptive (Toye et al. 2014). The data that form the basis of the meta-ethnography are not those collected through interviews, but are the authors' interpretations and explanations of these findings (Noblit & Hare 1988). These authors' interpretations therefore become the data for the synthesis (figure 12). These are recorded in either the authors' own words, paraphrased or summarised. Through reading, re-reading and remaining focussed on the aim of the synthesis the concepts for each study began to emerge. In Microsoft Excel, a worksheet for each study was created, in this I recorded the concepts identified in each study and the authors' interpretations that determined them. I also began to record my views and interpretations of these, an example is given in table 12.

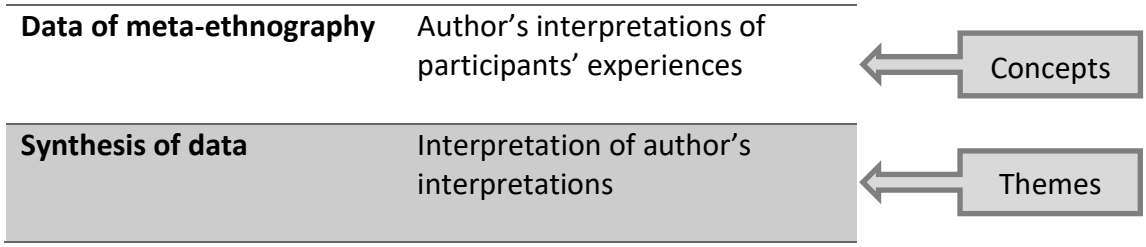


**Figure 12 Terminology used in meta-ethnography**

**Table 12 Example of synthesis of study data**

| concepts from paper 2 | interpretations from authors | *illustrative quotes* | *thoughts* | *concepts* |
|---|---|---|---|---|
| learning to live with diabetes | diabetes was a nuisance | | | *diabetes a nuisance* |
| | attitude of having to get on with their lives and 'deal with it' | *right I can deal with another thing, that's fine...and I just coped with it...and, yeah, I got positive about it. Although it was an extra hassle, I could deal with it* | *life balance* | *trying to live life* |
| | despite 'expert patients', highly experienced in management of CF, not equipped with knowledge & confidence for managing CFD | | *lack knowledge & confidence CFD* | |
| | commencing CFD treatment was quickly associated with symptomatic improvements- facilitated acceptance | *When you have treatment and feel much better that obviously, you know, makes you feel much happier about things* | | |
| | one of biggest barrier in the early stages of adjustment to diabetes was injecting insulin | | *insulin barrier initially* | |
| | apply CF coping strategies, lifestyle patterns & attitudes to the new challenge of managing diabetes | | *learning from CF behaviours helps* | *CF helps mange CFD* |
| | positive problem solving attitude to diabetes did not immediately guarantee good glycaemic control | *I think I cope well in terms of my attitude and so on, you know—I don't get down about stuff. But, umm, yeah, on a practical level I think I've made a lot more mistakes with diabetes in terms of...my sugar levels being too high* | | |
| | lack energy associated with CF made it difficult for some to live with diabetes | | *CF hinders management of CFD* | *Cf hinders CFD mangement* |
| | conflicting demands of CF | *If I didn't have cystic fibrosis and I was just diabetic, and I didn't feel so shitty all the time, then I probably could do it, but because I find my CF sort of takes over,* | *conflicts of CF* | |

In meta-ethnography, there is more than one level of interpretation. Schutz's (1962) notion of first, second and third order constructs have been adopted in the analysis process by some meta-ethnographers (Britten et al. 2002, Campbell et al. 2003, Malpass et al. 2009). Where, first order constructs represent the everyday understandings of ordinary people (patient views), second order constructs refer to the constructs of social science (the author's views and interpretations) and third order constructs interpret findings beyond the original study (interpretations of synthesis team) (Britten et al. 2002, Malpass et al. 2009).

The data of meta-ethnography are what Schutz refers to as second-order constructs (Toye et al. 2014). However, the difference between first and second order constructs is not always clear for example authors may use participants' quotes as part of their interpretations. In the published studies authors, have selected the data they present and are more likely to choose participants' quotes/ experiences as 'good examples' of their interpretations and this therefore is not a representation of the full data-set of participants. The use of author's interpretations is thought to be more insightful as they propose an explanation of the observed phenomena (Atkins et al. 2008). Atkins (2008) suggests that all reported data are in fact the subject of author interpretations and therefore the usefulness of Schutz notions is unclear. Therefore, due to the lack of clarity with regards to the classification of orders of constructs and their benefit this meta-ethnography uses the authors' interpretations as its data source. If the notion of Schutz constructs were applied to the terminology used in this thesis (figure 12) second order constructs would be the same as concepts and third order constructs the equivalent to themes.

The concepts identified for each study were recorded alongside the authors' interpretations that determined them. Views and interpretations that started to arise as part of the data analysis were also noted.

### 3.1.5 Stage 4: Determining how the studies are related

The relationships between these studies were explored to ascertain how they were related. Similarities within the studies included: participant characteristics because they were all adolescents or adults with CFD and the concepts identified within the studies e.g. CF is worse, CFD is intrusive, CFD is part of CF. Therefore reciprocal translation of each study into each of the other studies could be conducted (Noblit & Hare 1988). From this reciprocal translation, a line of argument was developed.

### 3.1.6 Stage 5: Translating the studies into one another

Through repeated reading of the studies, common and recurring concepts were identified. Noblit and Hare (1988) do not give clear guidelines how to conduct this, therefore the method of Atkins et al (2008) was followed. The studies were arranged

into chronological order and the concepts of paper one were compared with paper two. The synthesis of papers one and two was then compared with paper three, concepts from this synthesis were compared with paper four and so on until all the studies had been translated into each other.

The synthesis may be influenced by the order in which the studies are compared (Atkins et al. 2008). Some authors suggest starting with a key paper by an expert within that field (Campbell et al. 2003). For this synthesis, such a paper or key author does not exist as the pool of studies was small. This synthesis was therefore carried out in chronological order as described by Atkins (2008). There were however only eight years between the publication dates of the studies therefore the impact of chronological order is likely to be insignificant.

### 3.1.7 Stage 6: Synthesising translations

The identified concepts were written on index cards and through the process of constant comparison between each concept they were placed into groups which had a common meaning (figure 13). Through interpretation, asking questions such as what does this mean? does this impact on another group or it meaningful on its own? what is the relationship? these groups were developed into themes to reflect these shared meanings (SC).

**Figure 13 Process of constant comparison**

The research team (SC, JS, SW, AJ) explored all the themes and the rationale behind them, and challenges to interpretations were reflected upon. Themes were reworked upon reflection (SC), reading and re-reading the studies until it was felt that they best represented all the identified concepts.

These themes were then looked at holistically to develop a line of argument synthesis (SC, JS, SW, AJ). This is the development of an interpretation – the discovery of a whole amongst a set of parts (Noblit & Hare, 1988). It represented the overarching perspective of adolescents and adults in living with and managing CFD. A visual representation was then developed to reflect the line of argument synthesis.

### 3.1.8 Stage 7: Expressing the synthesis

This stage involves dissemination of the findings for maximal impact. The synthesis is presented in the results and discussion sections that follow.

There are currently no reporting guidelines for meta-ethnography; these are currently being developed and will address methodological and reporting standards to maximise the quality of meta-ethnographical research and its contribution to health services and patient care (France et al. 2015). ENTREQ guidelines for improving transparency in qualitative evidence synthesis are available (Tong et al. 2012). However, these are generic and are not being widely used to inform meta-ethnography reporting as they are not designed for the distinct synthesis process of meta-ethnography (France et al. 2015).

### 3.1.9 Quality in qualitative evidence synthesis

To address quality this meta-ethnography aims to demonstrate transparency in the reporting of its processes of data collection and analysis. Strengths and limitations of the study will also be explored and reflected upon.

#### 3.1.9.1 Reflexivity

Given that this meta-ethnography is concerned with the synthesis and interpretation of qualitative research, it is important to be transparent about the researcher and their knowledge, experiences and assumptions. As a healthcare professional, with over 20

years of experience working with adults with CFD, I have a vast knowledge of what I believe CFD management involves. However I am mindful that I have only ever worked in one CF centre and that practices between CF centres and individual patients can vary. I will therefore remain open to new insights and experiences. Findings and interpretations will also be explored with the research team to ensure they are true reflections of the data. I am also the lead author of one of the identified studies but as meta-ethnography does not involve the re-analysis of raw data but the synthesis of author's interpretations of participant's experiences this was not regarded as a conflict of interest.

## 3.2 Results

### 3.2.1 Appraisal of included studies

This meta-ethnography synthesised qualitative research studies concerning adolescents and adults with CF experiences of living with and managing CFD.

A total of 1584 records were identified after duplicates were removed (figure 14). The title and abstracts for each record were screened by a single author to identify qualitative studies which addressed the research question "what are the experiences of adolescents and adults with CF in living with and managing CFD?" Full text articles were read if these criteria were met or there were uncertainties in methodology (n=39). Six qualitative studies were identified for inclusion in the synthesis.

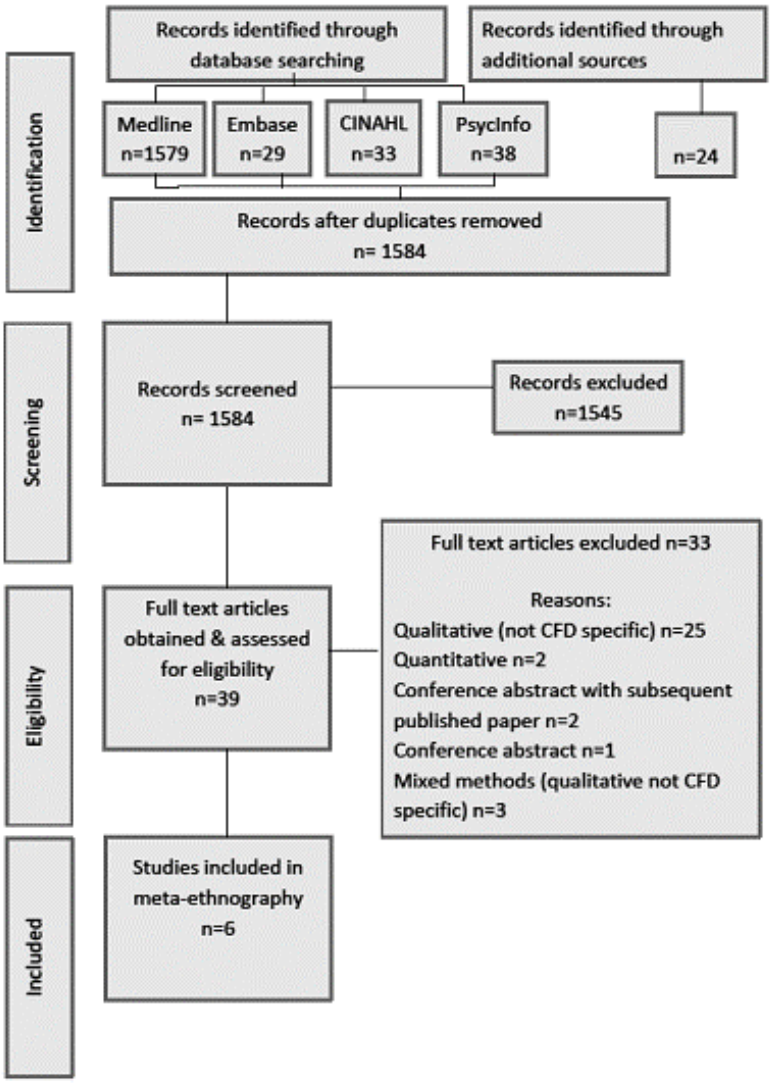


**Figure 14 Flowchart of study selection process**

All six included studies were appraised using the modified CASP criteria (Campbell et al. 2003), this consisted of two pre-appraisal questions followed by 10 standard CASP questions which focussed on validity, results and clinical relevance (table 13). All identified studies met the pre-appraisal questions and were therefore further appraised according to CASP criteria. When appraising a study using CASP there is the option of yes, no or can't tell answers. Table 13 demonstrates that most of the CASP criteria was met by the majority of included studies. No studies were excluded as a result of this critical appraisal. There is debate about whether including a poorer quality study in an evidence synthesis would be detrimental as these studies could be

rich in data (Campbell et al. 2011). However, in this meta-ethnography no studies were excluded based on quality; they were excluded if they didn't report qualitative findings and they had insufficient focus on CFD.

**Table 13 Modified CASP criteria**

| Pre-appraisal question | Yes | no | |
|---|---|---|---|
| | Paper number | | |
| Does the study report results from qualitative research and did it involve both qualitative methods of data analysis and collection? | 1,2,3,4,5,6 | | |
| Is this research applicable to the synthesis topic? | 1,2,3,4,5,6 | | |
| CASP screening questions | yes | no | Can't tell |
| | Paper number | | |
| Was there a clear statement of the aims of the research? | 1,2,3,4,5,6 | | |
| Is a qualitative methodology appropriate? | 1,2,3,4,5,6 | | |
| Was the research design appropriate to address the aims of the research? | 1,2,3,5,6 | | 4 |
| Was the recruitment strategy appropriate to the aims of the research? | 1,2,3,4,5,6 | | |
| Was the data collected in a way that addressed the research issues? | 1,2,3,4,5,6 | | |
| Has a relationship between researcher and participants been adequately considered? | 1,3,6 | 4 | 2,5 |
| Have ethical issues been taken into consideration? | 1,2,3,4,5,6 | | |
| Was the data analysis sufficiently rigorous? | 1,2,3,5,6 | | 4 |
| Is there a clear statement of findings? | 1,2,3,4,5,6 | | |
| How valuable is the research?<br>*Contribution of study to existing knowledge or understanding*<br>*Identify new areas of research*<br>*Translations of findings to other populations or other ways research used* | <br>1,2,3,4,5,6<br><br>1,2 ,3,4,6<br>2,3,4,6 | <br><br><br>5<br>1,5 | |

Table 14 summarises the key characteristics of the six studies included in this meta-ethnography, this includes: country of origin, aims, method of data collection and

analysis and participant details (age, gender and where available treatment of CFD). This meta-ethnography is a collective representation of 83 adolescents and adults with CFD from three different countries. The three earlier studies were conducted in the UK (Collins & Reynolds 2008, Tierney et al. 2008, Richmond 2012) with the remaining three being conducted in the USA or Canada (Dashiff et al. 2013, Millington et al. 2014, Sylvain et al. 2016). It is important to consider the setting of the studies as there are differences in healthcare provision between these three countries. All studies used qualitative methodology involving interviews as the source of data collection. In the mixed-methods studies (Dashiff et al. 2013, Millington et al. 2014) the quantitative results were excluded from the synthesis. Only data from adolescents or adults with CFD were included in the synthesis, accounts from parents (Millington et al. 2014) and those with normal glycaemia or impaired glucose tolerance (Sylvain et al. 2016) were excluded. This is because the focus of this study is on the experience of living with and managing CFD. It was not clear in some of the studies what treatment all the participants were receiving (Richmond 2012, Sylvain et al. 2016). However most of the treatments alluded to by the participants tended to focus on the use of insulin. Where there was uncertainty it was highlighted as part of the interpretative process.

**Table 14 Summary of included studies**

| Paper number Authors and year | Country of study | Aims | Method | Participants |
|---|---|---|---|---|
| **Paper 1**<br><br>**Tierney S et al. 2008** | UK | To explore the views of people with CFD and type 1 diabetes on the diagnosis and management of diabetes | Semi-structured interviews (face-to-face or telephone)<br><br>Framework analysis was used to analyse the data | 11 adults with CFD, mean age 30.4 years<br>6 female<br>all on insulin injections<br><br>Did not include analysis from people with type 1 diabetes |
| **Paper 2**<br><br>**Collins S and Reynolds F 2008** | UK | To examine the experiences of adults with CFD in adapting to the diagnosis of diabetes | Semi-structured face-to-face interviews<br><br>Interpretative phenomenological analysis (IPA) was used to analyse the data | 22 adults with CFD, mean age 34 years<br>10 female<br>20 on insulin injections<br>1 on oral medication |
| **Paper 3**<br><br>**Richmond R 2012** | UK | To evaluate a structured dietary education programme for patients with CFD | Semi-structured face-to-face interviews<br><br>Framework analysis was used to analyse the data | 8 adults with CFD, mean age 30.9 years<br>5 female<br><br>Treatment- not stated |

| | | | | |
|---|---|---|---|---|
| **Paper 4**<br><br>**Dashiff C et al. 2103** | USA | To describe parents' and older adolescents; experiences and perceptions of the diagnosis of CFD and its management.<br><br>To describe parents; and adolescents' perceptions of parental behaviour that supported self-management. | Mixed methods- semi-structured interviews were used to capture participants' perceptions<br><br>Questionnaire to measure parental support and autonomy.<br><br>Qualitative analysis as described by Sandelowski | 10 adolescents with CFD, mean age 16.6 years<br>3 female<br>9 on insulin injections<br>1 on insulin pump<br><br>Only adolescents' experiences and perceptions from the qualitative interviews were included. |
| **Paper 5**<br><br>**Millington K et al 2014** | USA | To examine the reactions of children and adolescents, their parents, and adults with CFD to CFD management and factors that facilitate or impede treatment adherence | Mixed-methods- semi structured interviews (face-to-face or telephone) and CF quality of life questionnaire<br><br>Phenomenological analysis used to analyse the interview data | Ten children and adolescents (mean age 14.6 years) with CFD, 3 female<br>1 participant on insulin pump, 9 insulin injections<br><br>10 adults with CFD (mean age 29.5 years), 3 female<br>3 on insulin pumps, 7 insulin injections<br><br>10 parents<br><br>Only accounts of adolescents and adults with CFD were included. |
| **Paper 6**<br><br>**Sylvain C et al. 2016** | Canada | To explore patients' representations of CFD to better understand the discrepancy between patients' expected and observed health behaviours | Semi-structured face to face interviews<br><br>Qualitative data methods (Miles, Huberman & Saldana, 2014)- thematisation was structured around Leventhal's five dimensions of illness representation | 22 adults with CFD, mean age 30.6 years<br>16 female<br><br>Only adults with CFD included (not those without dysglycaemia or those with IGT)<br><br>Treatment- not stated |

### 3.2.2 Synthesis findings

There is a degree of overlap between the processes of reading, deciding how the studies are related and translating one study into another, therefore these results are presented together. Table 15 highlights how concepts were initially identified in one of the studies through the processes of repeated reading. Throughout the process of translation, common and recurring concepts were identified in all the studies, these are summarised in table 16. These identified concepts were then synthesised into themes to reflect their shared meaning (table 17).

## Table 15 Identification of concepts in one study

| | main findings from paper | Theme (author's interpretations) | *concepts 3* |
|---|---|---|---|
| R Richmond | | grown up with CF so learned to live with it | *CF is the normal* |
| 2012 | coming to terms with diagnosis | generally found CFD much harder to deal with (regardless number years since diagnosis & social support) | *CFD is hard to deal with* |
| | | difficulty in accepting diagnosis "getting their head around it" or saying took a long term "to come to terms with it" | |
| | | negative feelings at this time | |
| | | diagnosis more significant for some patients than others | *is an individual varaition to how see CFD* |
| | | how they had tried to ignore it and hoped it "would go away" | |
| | | The impact of CFD on patients may be underestimated and should be recognised more by the CF team so that support can be given to patients to help with accepting it | *Acceptance of CFD will facilitate management* |
| | | | |
| | | more intrusive than CF | |
| | | CF worse than CFD ***one patiet | *CF is worse* |
| | | rather than seeing CFD & its treatment as a 2nd chronic disease which required treatment it was often reported as being just something else that patients had to deal with on top of CF treatment | *CFD part of CF* |
| | living with the constancy of CFD | CFD was more difficult to manage than CF, required more continual, ongpoing input than managing CF. | *can't hide behind CFD* |
| | | CFD did not "fade into the background" in the same way that CF did | *CFD needs constant attention - increases treatment burden* |
| | | CF treatment could be completed at home & behind closed doors | |
| | | CFD treatment could not be "hidden" in the same way as CF treatment | *CFD is always there* |
| | | constancy of CFD felt by patients and increased treatment burden to be completed alongside CF treatment - length of time been diagnosed did not effect this | |
| | | need for extra thought about CFD- particularly before meals | |
| | | the work needed to manage it & the forward planning to help control their CFD, dependent on what they were going to do next | |
| | | more intrusive than treatment for CF | *more intrusive than CF* |
| | | CFD treatment described as being continual throughout the day with the need for monitoring BG levels & injecting insulin also the need for extra thought about CFD- particularly before meals when deciding on insulin they would ned to give for the food required | *CFD is always there- requires constant attention* |
| | | | |
| | patients' perspectives on the mangement of CFD | monitoring of BG levels as being on of the most intrusive parts of managing CFD | *CFD more intrusive* |
| | BG monitoring | constant monitoring' and the difficulty trying to remember the correct time when they were busy with other things or working | |
| | | increasing frustration at having to do the monitoring was reported as being one of the worst parts of managing CFD, resulting in lack of motivation to monitor BG levels amongst some patients | *CFD needs continual attention, can't hide from italso need to forward plan* |

**Table 16 Summary of concepts identified in each paper**

| Authors and year<br>Paper number | Main themes from papers | Summary of author's interpretations | Concepts |
|---|---|---|---|
| Paper 1<br><br>Tierney S et al. 2008 | • An evolving vs fractured identity<br>• Balancing<br>• Motivation<br>• Isolation | *Conflicts occur between CF and diabetes management*<br>*Considerations needs to be given to how people with CF view diabetes, some view it as part of CF. This may influence its management.*<br>*Having CFD makes CF visible.*<br>*There is a sense of trying to manage in 'normal CF life'*<br>*CF life limiting*<br>*CFD associated with a deterioration in health*<br>*Having CF is seen as both a help and hindrance to CFD management*<br>*Having diabetes can help and hinder CF management* | *Is CFD part of CF?*<br>*CF more important*<br>*Living with CFD*<br>*Learning to live a new normal*<br>*CFD makes CF visible*<br>*Try to live 'normal' life*<br>*CF life limiting*<br>*Health deterioration, progression of primary disease*<br>*CF can help or hinder CFD management*<br><br>*CFD can help or hinder CF management* |
| Paper 2<br><br>Collins S and Reynolds F 2008 | • Emotional response to diagnosis<br>• Looking for an understanding<br>• Learning to live with diabetes<br>• Limiting the impact of the diagnosis | *initial feelings about diabetes shaped by experiences with CF*<br>*Diabetes was a nuisance*<br>*confront difficult feelings about CF, which may have seemed well-controlled & normal*<br>*Conflicts occur between CF and diabetes management*<br><br>*CFD perceived as an additional practical demand on their health-management strategies*<br>*Apply CF coping strategies, lifestyle patterns & attitudes to the new challenge of managing diabetes*<br>*lack energy associated with CF made it difficult for some to live with diabetes*<br>*not as significant as living with CF* | *Is CFD part of CF?*<br><br>*CFD is a nuisance*<br>*CFD makes CF visible*<br><br>*How they see CFD influences how they manage, do you have to accept the diagnosis to manage well?*<br>*Increase anxiety regarding CF*<br>*Trying to manage in 'normal CF life'*<br>*CFD requires additional more practical demands/attention*<br>*Having CF helps/hinders CFD management*<br><br>*CF more important* |

| | | | |
|---|---|---|---|
| | | *Difficulty and uncertainties in balancing CFD particularly diet and blood glucose*<br>*Diabetes coincided with serious worsening of CF in some provoking feelings of powerlessness and depression*<br>*Knowledge and support are key to managing CFD- this is relevant to patients, healthcare professional and peers/ family.* | *Control and balance*<br><br>*Impact on health*<br><br><br>*Knowledge and support- requirements, for who* |
| Paper 3<br><br>Richmond R 2012 | • Patient's perspective on the impact of CFD<br>  ○ Coming to terms with the diagnosis<br>  ○ Living with the constancy of CFD<br>• Patient's perspectives on the management of CFD<br>  ○ Blood glucose monitoring<br>  ○ Administering insulin injections<br>  ○ Lifestyle considerations and flexible treatment regimes | *There is an individual variation to how participants see having CFD.*<br>Generally, found CFD much harder to deal with as they grew up living with CF<br>*Acceptance of CFD was seen to facilitate management*<br>Constancy of CFD felt by patients and increased treatment burden to be completed alongside CF treatment<br>*You can't hide from CFD, it also needs to forward plan.*<br><br>*Lack of CFD control impacts on CF health.*<br>*Knowledge is key to managing CFD.* | *Individual view of CFD*<br><br>*CFD is hard to deal with*<br>*CF is the normal*<br>*Acceptance of CFD will facilitate management*<br><br>*CFD need continual attention- increases treatment burden*<br>*CFD is always there*<br><br>*Impact on CF health*<br>*Knowledge is key* |
| Paper 4<br><br>Dashiff C et al. 2103 | • Adolescents' experiences and understanding of CFD<br>• Adolescents' perceptions of their self-management of CF and CFD<br>• Adolescents' perceptions of parental autonomy support | *Conflicts with other types of diabetes can help or hinder the management of CFD*<br>*Having CFD increases the treatment burden and requires more practical demands*<br>*Diabetes needs looking after but it can be difficult to deal with diabetes.*<br>*CFD management becomes part of CF routine/adaptation.*<br>*Having CFD can promotes CF care*<br>*Parents help support adolescents with disease management tasks.* | *Help or hindrance from other forms of diabetes*<br><br>*Increase burden of treatment*<br><br>*Need to look after CFD*<br>*CFD difficult to deal with*<br>*CFD becomes part of CF routine*<br><br>*CFD promotes CF care*<br>*Parental help (adolescents)* |

| | | | |
|---|---|---|---|
| | | *Knowledge and ongoing support are key to managing CFD- this is relevant to patients, healthcare professionals and peers/ family* | *Knowledge and ongoing support key for all involved* |
| Paper 5<br><br>Millington K et al 2014 | • Prior knowledge of diabetes<br>• Reactions to CFD management<br>• Barriers to CFD treatment adherence<br>• Factors promoting CFD treatment adherence<br>• Patient responsibility for CFD management<br>• Role of the healthcare team | *Screening for CFD helps increase the awareness of the possibility of developing CFD.*<br>*Having CFD increases the treatment burden and requires more practical demands and attention*<br>*the addition of CFD management is a risk factor for compromised adherence to both regimens*<br>*Management of CFD needs to be part of routine CF care.*<br>*Having CFD makes CF visible.*<br>*CF more important than CFD*<br>*Having CF is seen as both a help and a hindrance CFD management.*<br>*the diagnosis of CFD signifies a major disruption to their 'normal life'*<br>*Present CFD represents a significant disruption*<br>*Lack of CFD control impacts on CF health.*<br>*CFD management needs to become part of CF routine/adaptation.*<br>*Parents help support adolescents with disease management tasks*<br>*Diabetes needs looking after but it can be difficult to deal with and impact on daily life.*<br>*Knowledge and support from healthcare professionals are key to managing CFD-* | *Screening increases awareness CFD*<br><br>*Increase burden of treatment*<br>*Living with CFD increases treatment burden*<br>*Additional treatments compromise adherence*<br><br>*CFD becomes part of CF routine*<br><br>*CFD makes CF visible*<br>*CF more important*<br>*CF can help or hinder CFD management*<br><br>*CFD disturbs life*<br>*Learning to live a new 'normal' life*<br><br>*Controlling CFD affects health*<br>*Part of routine CF care*<br><br>*Parental help (adolescents)*<br><br>*Influence of CFD on CF health*<br><br>*Knowledge is key to management* |
| Paper 6<br><br>Sylvain 2016 | • Identity<br>• Cause<br>• Timeline<br>• Consequence | *CF more important than CFD as CF is a life-limiting disease.*<br>*However, there is acknowledgement that diabetic complications can occur.*<br>*patients tended to view CFD as a permanent problem* | *CF is life-limiting*<br>*CF more important*<br>*Diabetic complications can occur*<br>*CFD permanent*<br>*Impact on QoL* |

| | | | |
|---|---|---|---|
| | • Control<br>• Knowledge | *Quality of life is important - need to strike a balance with doing treatment and quality of life. Patients choose to selectively non-adhere on occasions.*<br>*Th*e rigorous treatment of diabetes coming second<br>*CFD is not thought of as being the same as other forms of diabetes.*<br>*There is a close link between CF and CFD but conflict if CFD is part of CF.*<br>*Influence on CFD management from those close to the patients, such as family, friends and work colleagues.,*<br>*Knowledge of healthcare professionals was essential to patients, they reported a lack of knowledge and a conflict in knowledge about CFD amongst cares and families*<br>*CFD representation is intertwined within the experience of living with CF.* | *Selective non-adherence*<br><br>*Treatment CFD not always important*<br>*CFD different from other diabetes*<br><br>*CFD is a distinct disease*<br>*CFD is part of CF*<br><br>*Influences outside the care system*<br><br>*Knowledge is key*<br>*Lack knowledge carers/ families*<br>*Conflicts in knowledge*<br>*CF and CFD intertwined* |

**Table 17 Synthesis of identified concepts into themes**

| concepts 1 | concepts 2 | concepts 3 | concepts 4 | concepts 5 | concepts 6 | theme |
|---|---|---|---|---|---|---|
| is CFD part of CF<br>CF more important | progression of CF<br>CFD is a nuisance<br>is CFD part of CF | Acceptance of CFD will facilitate management | conflicts with other types of diabetes can help or hinder | not as significant as CF<br>screening increases awareness<br>CF-CFD interaction<br>CF affects CFD control<br>uniqueness of CFD- | CFD is not the same as other diabetes<br>CFD is not static<br>CFD evolution of CF<br>CF and CFD intertwined - underregulation and misregulation'<br>CFD consequence of CF treatment<br>close link between CF and CFD but conflict if CFD is part of CF<br>cf more important, impact on survival | A need to develop a perspective on CFD |
| CFD makes CF visible | how they see CFD influences how they manage, do you have to accept the diagnosis to manage well?<br>CFD makes CF visible<br>CFD increases anxiety about CF | is an individual variation to how see CFD<br>can't hide behind CFD | | making CF visible<br>CFD is hard to deal with, disturbs life<br>screening helps prepare | CF and CFD representations intertwined with experience living with CF<br>CFD distinct disease- separate treatment<br>permanent vs intermittent | struggling with identity |
| CF life limiting<br>health deterioration, progressive not fractured-progression of primary disease | illness beliefs effect on health<br>CF life limiting<br>understand and recognise convergence-key to control health | lack of CFD control impacts health | need to look after diabetes | Controlling CFD = better health | complications occur | A representation of morbidity and mortality |

| | | | | | | |
|---|---|---|---|---|---|---|
| *diabetes can help and hinder CF management*<br><br>*trying to manage 'normal CF life'*<br><br>*CF helps/hinders CFD management* | *CFD requires additional more practical demands/attention*<br><br>*trying to manage in 'normal CF life'*<br><br>*more to do - do they balance this*<br><br>*CF heps/hinders CFD management* | *CFD is always there*<br><br>*age and partners may influence some*<br><br>*CFD needs constant attention - increases treatment burden*<br><br>*CFD needs continual attention, can't hide from it also need to forward plan* | *increase treatment burden*<br><br>*difficult to deal with diabetes*<br><br>*promotes CF care becomes part of CF routine/adaptation* | dietary concerns<br><br>learning to live with a new 'normal'-<br><br>more to do!<br><br>CFD becomes part of CF routine- helps<br>*CFD management needs to be part of routine CF care*<br>BG testing one of biggest challenges<br>having CF helps<br>forgetting -common reason for non-adherence<br>pain of injections and BG monitoring<br>more treatments impact adherence | *comprehensive and integrated care, flexible to meet changes in health*<br>*QoL important, Cf more important than CFD, treatment CFDnot always important*<br>*influences outside the care system/ tensions in daily life* | **in control or being controllled** |
| *knowledge and support are the power (own/peers/HCP/public)* | *knowledge and support are the power (own/peers/HCP/public)* | *knowledge is key* | *lack confidence/skills*<br><br>*knowledge is power*<br><br>*parental help (adolescents)*<br><br>*support ongoing CFD education programmes needed* | confusion re diet<br>*knowledge and experience of managing from health care teams required*<br><br>*knowledge is power*<br>HCP need knowledge of all aspects of CF and CFD care (including CFD and diet)<br><br>*parental help (adolescents)* | *knowledge- CF team key*<br><br>*lack of knowledge carers/ families/ conflicts*<br>*knowledge/diet*<br>*knowing does not mean doing with regards to insulin and BG checking* | **need for knowledge and support** |

**Table 18 Identified themes**

| Theme |
|---|
| A need to develop a perspective on CFD |
| Struggling with identity |
| A representation of morbidity and mortality |
| In control or being controlled |
| Need for knowledge and support |

Five themes emerged from synthesising translations. These are: a need to develop a perspective on CFD, struggling with identity, a representation of morbidity and mortality, in control or being controlled and need for knowledge and support (table 18). These themes acknowledge the core experiences that influenced the management of CFD, with the central phenomena concerning how to achieve a sense of balance between CF and CFD. With the exception of Dashiff et al. (2013), which did not have concepts relating to the theme struggling with identity, all papers had concepts relating to all the identified themes (table 19). A detailed description of each of these themes are presented below.

**Table 19 Occurrence of themes**

| Theme | **Paper 1** Tierney S et al. 2008 | **Paper 2** Collins & Reynolds 2008 | **Paper 3** Richmond R 2012 | **Paper 4** Dashiff C et al. 2103 | **Paper 5** Millington K et al 2014 | **Paper 6** Sylvain 2016 |
|---|---|---|---|---|---|---|
| **A need to develop a perspective on CFD** | √ | √ | √ | √ | √ | √ |
| **Struggling with identity** | √ | √ | √ | | √ | √ |
| **A representation of morbidity & mortality** | √ | √ | √ | √ | √ | √ |
| **In control or being controlled** | √ | √ | √ | √ | √ | √ |
| **Need for knowledge & support** | √ | √ | √ | √ | √ | √ |

For the ease of reading in the following section the papers will be referred to by numbers (as identified in table 19 rather than authors).

#### 3.2.2.1 A need to develop a perspective on CFD

How CF is viewed and managed influences individual's thoughts and feelings about CFD (2). CFD needs to be managed in a world of competing treatment demands and pressures associated with managing CF, with most seeing CF as more important to them (1,2, 5, 6).

> *If I didn't have cystic fibrosis and I was just diabetic, and I didn't feel so shitty all the time, then I probably could do it, but because I find my CF sort of takes over, and that makes me feel ill and there's stuff I can't do and I can't cook my dinners and…I want to just lie in bed and feel like I'm dying. Then my diabetes, you know, just kind of takes a second…thing. (2)*
>
> *it's been a pain with the insulin and stuff…but the CF's worse than the diabetes. (3)*

There were conflicts in perceptions as to whether CFD was part of CF or a distinct entity. Despite some recognising the close link between CF and CFD they sometimes considered it a distinct disease treated separately by a different doctor and additional treatments (1, 6). The representations of CFD were described as intertwined with the experience of living with CF (6). For others it is difficult to separate the two conditions which both directly impact upon each other.

> *I just kind of treat it purely as a weight thing rather than a separate disease. (1)*
>
> *As long as I see diabetes as a secondary disease, I can't put the two together and compare them like that. It's like they're two different things even though they go with the same health problem'. (6)*

Being diagnosed with CFD provoked mixed emotions and was more significant for some. Many reported negative feelings at the time of diagnosis (2-5), which included descriptions such as 'shocking,' 'bad,' 'hard,' 'terrible,' 'depressing' and 'agitating '(4). One study found that CFD was generally regarded as much harder to deal with in terms of control, acceptance and impact on daily life and it took time "to come to terms with it" (3). Conversely having CFD was also seen as easy and "not a big thing" (4). A relief at the diagnosis of CFD was experienced by a few participants as it provided an

explanation of recent symptoms (2, 4). Routine annual CFD screening highlighted the possibility of being diagnosed with CFD and helped acceptance when diagnosed (5).

The experience of having CF helped people come to terms with the diagnosis and management of CFD by applying existing lifestyle patterns, routines, coping mechanisms and strategies (2, 5) and through being accustomed to living with a chronic illness (1). There are lots of uncertainties from living with an unpredictable long-term condition like CF (6). One of the biggest uncertainties experienced in managing CFD were with regards to managing diet and CFD, where potential or recommended dietary changes were seen as distressing for some (5) and no changes were met with a sense of relief (1). Difficulties were seen in balancing diet and controlling blood glucose (2). Dietary differences were also the main dissimilarities highlighted between CFD being different to type 1 or type 2 diabetes (1,5,6).

> *If I didn't have CF and weight wasn't such an issue, the sugar side of the diabetes wouldn't be so bad because I'd, I could just eat a healthy diet and not worry too much about…trying to eat a lot of food with the CF side of things and trying to keep my sugars balanced as well. You know, it's very difficult to do both. (2)*

#### 3.2.2.2 Struggling with identity

CFD did not "fade into the background" in the same way that CF did; its treatments could not be 'hidden' the same way as CF treatments (3), people had to deal with managing their health in public (5), for example checking blood glucose levels or administering insulin injections (3, 5). Therefore, for many the development of CFD made CF visible. This disrupted *'normal'* identity, interfered with lifestyle (5) and brought feelings about CF to the foreground (2). However, managing CFD did not define their life and they tried to live life as 'normally' as they could despite having CFD (1).

> *Cus like most of my treatment for that (CF) as well can be done at home behind closed doors and everything else but with that (CFD), you don't, or many times you don't have a choice but to do it out and about and even in today's day and age, people have got issues with it. Even work colleagues and everything else have issues with you doing injections which I used to think well if they've got an*

*issue with it, I'll just leave it or whatever but I've come to the conclusion now if they've got an issue with it, that's their problem. (3)*

*I saw it as something that would hamper my lifestyle. Something that would, you know, interfere… for the most part until then I just had to worry about taking some enzymes maybe an antibiotic. During the day it didn't really impact me…other than a few little things here or there I was living 'a normal life.' So, that was kind of a major hurdle major for me. (5)*

#### 3.2.2.3 A representation of morbidity & mortality

A lack of glycaemic control had negative impact on weight, lung function and overall health (1, 3, 5, 6). When unwell with CF-related illnesses, such as respiratory exacerbations, it was more difficult and challenging to control blood glucose levels due to the increased demands made upon self-management (1, 5), lack of energy (2) and the increase in and impact of some CF treatments, noticeably corticosteroids (5). For some the development of diabetes marked the point when their CF health started to deteriorate, or they worried their health was going to deteriorate (1, 2).

*It was something that you didn't want to accept because it's an acceptance of the disease progressing … I had to wrestle with the fact that it was a progression of the CF. (1)*

*As my health dropped, as it did at that point because my sugars were so high, I suddenly became prone to infection…I was in Hospital X for about 4 weeks. I was really quite poorly, I was really quite fed up, so there was only two times in my life I'd ever say I was clinically depressed…I think that was one of the times. (2)*

*The CF, it does what it wants to do really. So you just have to deal with it rather than prevent it. With the insulin, you're avoiding a problem; with the CF you've got to deal with a problem once it's happened … especially as I'm getting older. You do what you can but things happen. (1)*

In terms of the overall impact on health CF was regarded as worse because it was considered life-limiting (1, 2, 5). Although the impact of the development of CFD on morbidity and mortality were acknowledged (5), the possibility of diabetes related complications was not of great concern for health (1, 6) with the likelihood of succumbing to CF first being greater (1).

*I know about the long-term consequences, blindness, the circulatory problems, the kidney disease, bla bla bla. I have to say that my viewpoint is … if the diabetes ever gets to the stage where it causes me those sort of problems I'll be dead chuffed because I'll still be here. I won't be happy that I'm suffering them, but I suspect the CF will get me first. (1)*

*I couldn't let my diabetes go any more just because the repercussions are long term. So now if I don't inject my insulin this evening, say, my glucose is going to go up, which could be dangerous long term … While if I take my treatment [for cystic fibrosis] this evening, I know I'm going to feel the effects right away. (6)*

#### 3.2.2.4 In control or being controlled

The development of CFD was associated with an increase in burden of treatment and monitoring (1-3, 5) which impacted upon all aspects of daily life (2, 3, 5). Diabetes had to be managed in a world which was already full of treatment demands and pressures (1). The addition of 'one more thing' to be completed alongside complex CF treatments was frustrating (2, 3). This compromised adherence to both CF and CFD regimens which influenced control in terms of CFD management, but also feelings of being in control (5). Incorporating CFD management into daily CF regimens (1, 5) and experience drawn from managing CF and its associated treatments helped achieve control of CFD and a sense of balance of disease management (4).

*Some people let it affect their whole life, whereas it doesn't need to. You shouldn't let it rule your life. You should try and fit your life in with it. (1)*

*Again I thought it was, oh another thing I've got to deal with, but at the time that was when…my CF was quite under control and it was just like, right I can deal with another thing, that's fine…and I just coped with it…and, yeah, I got positive about it. Although it was an extra hassle, I could deal with it. (2)*

*'I don't like it [insulin] but in the end it's really a very simple solution to control something that's really hard to control'. (6)*

CFD required more continual attention than CF, particularly monitoring blood glucose levels, administering insulin and making decisions about food (1, 4); its management required more forward planning (3). The constant attention of CFD management required treatments to be conducted outside the home. Monitoring blood glucose levels was one of the most intrusive parts of CFD management (3). A dislike and

embarrassment when giving insulin or monitoring blood glucose levels in public was experienced (3, 5). Remembering, lack of time, poor knowledge and pain were reasons for not monitoring blood glucose levels (1, 3, 5). Preserving quality of life was important and in order to maintain this CF treatments were often prioritised (1, 6).

> *You can't just go out and do what you want, when you want, you've got to think hard and plan it a bit better. It's inconvenient. (1)*

> *Even though all my glucose testing stuff is on my desk, in plain sight, it's like my brain doesn't see it in the same way it does my bottle of enzyme pills'. (6)*

> *So all the blood, erm checking my blood sugars and everything else, it's pretty much constant all the day and then if I'm doing sport when it comes to having my meals or whatever, I have to take into account like last week I had my tea and then I was going playing football so I had to balance it out with the insulin and everything else so if I'm being honest I'd probably say it has more of an effect than the CF. (3)*

#### 3.2.2.5 Need for knowledge and support

People with CF lacked knowledge and understanding about CFD (2-6); this was primarily related to what caused CFD and how to manage it. Many people with CF were not aware CFD was a potential complication of their CF (1-3, 5). There was also a lack of knowledge and clarity about CFD amongst friends, families and some healthcare professionals, such as general practitioners (1, 2). This was particularly related to dietary advice (2, 3, 5, 6). Where CFD was not understood by others a sense of despondency was expressed (1). Segregation, to minimise the risk of cross infection between individuals with CF, prevented face-to-face support from others with CFD (1, 3).

> *He's (GP) very up to date on the diabetic side but not in relation to CF so I spoke to him about it and he said … 'I don't understand how it relates to the CF', so I had to kind of go into that with him a little bit because he understood CF separately. He just wasn't aware of them together. (1)*

Most people received advice about CFD from their CF teams or specialist CFD services (3, 5, 6). There were difficulties into trying to access information regarding CFD in the public domain (1,3). Literature about diabetes was considered unhelpful as it failed to

address concerns specific to CFD, such as weight or lung function (1). Healthcare professionals need to ensure that people with CFD feel well supported and informed about this condition (1-3), due to its association with poor health outcomes. Healthcare professionals need to be aware that although these patients are experienced at managing CF it does not necessarily promote confidence to manage CFD (2,3). CFD management needs to be comprehensive, integrated into routine care, and adaptable to consider changes in health status (1, 2, 4-6).

### 3.2.3 Line of argument synthesis

Themes were considered holistically and developed into a line of argument synthesis. This is the development of an interpretation – the discovery of a whole amongst a set of parts (Noblit & Hare 1988). What emerged as important from this synthesis is how people perceive their CFD in the context of their CF. This synthesis demonstrates that people with CFD are struggling to put perspective on CFD and this influences how they view and manage the condition; is it viewed as part of CF or a separate entity? There was a difficulty in balancing the two conditions. The development of CFD challenged identity and how individuals saw themselves with illness where CFD brought illness into the foreground by making illness visible. Negative impact on health was observed but CF was regarded as more significant with difficulty balancing both treatment regimens. Knowledge deficits contributed to uncertainties in managing CFD with more support required. These are very negative findings that lead to poor disease management choices. New perspectives on CFD management are therefore needed.

The aim of synthesis of qualitative research is to achieve a level of understanding beyond that attained in the individual studies. This synthesis supports a line of argument that recommends for CFD management to be effective CFD needs to be considered and managed as part of CF. To conceptually represent this an integrated model of CFD management was developed (figure 15). This illustrates the complex nature of issues involved in balancing the management of CFD and CF. This model suggests CFD should be viewed and managed as part of CF. When considered as one condition CFD is managed as part of routine CF care; all treatments, therapies and self-care strategies such as behaviours, beliefs and coping mechanisms, are managed

together; what affects CFD will directly impact CF and vice versa. Incorporation of CFD as part of routine CF care resulted in better control and sense of balancing treatments and management. Regarding CFD as part of CF will help integrate identity and facilitate how individuals view CF. The emphasis needs to be placed upon CFD being a common complication of CF. Therefore, healthcare teams need to start thinking differently to facilitate this and develop services geared to help and support individuals to develop effective self-care strategies and integration of CFD management into routine CF care.

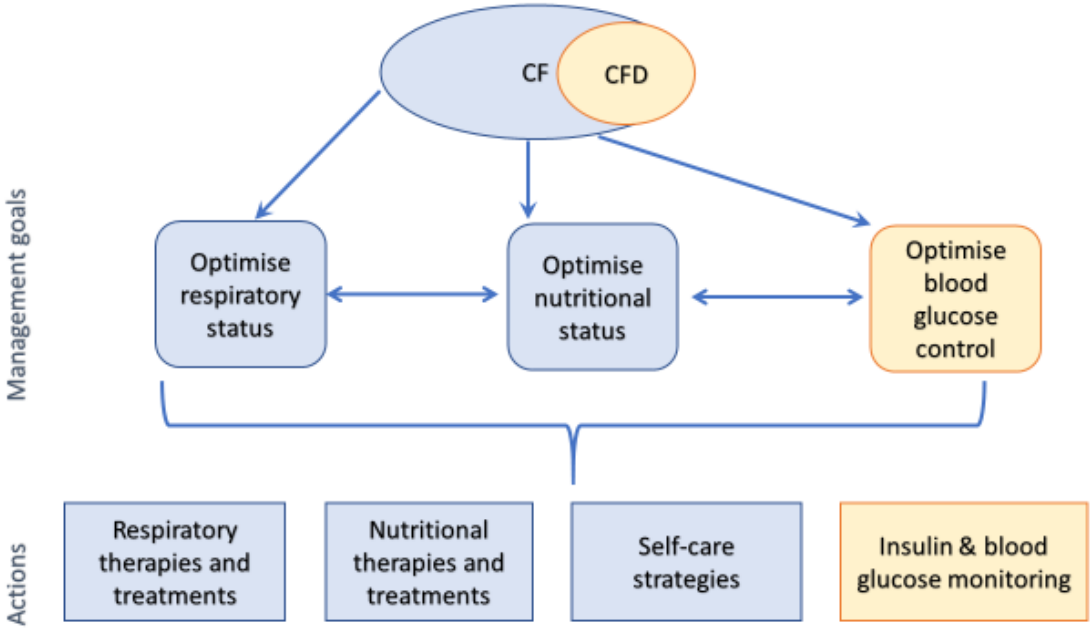


**Figure 15 Line of argument synthesis: striking the balance between CF and CFD**

## 3.3 Discussion

This meta-ethnography provides a greater understanding of how adolescents and adults with CF live with and experience CFD. It demonstrates that living with CF and CFD is a complex multi-dimensional process where the two conditions are closely intertwined. It identifies the key issue in responding to CFD is how CFD is viewed - is it part of CF or a separate entity? This directly impacts management because everything that is done to treat CF directly affects CFD and vice-versa. Living with CF and drawing upon the experiences developed in managing it were seen to help facilitate the

acceptance and management of CFD. Incorporating CFD management into CF regiments helped achieve a sense of control of CFD. An integrated model has been developed, it recommends CFD should be regarded as part of CF; it is a natural progression of CF that is seen with ageing. Due to the close links between nutritional status, lung function and diabetic control, managing CF and CFD as one condition will ultimately influence survival. It is not just about just treating the blood glucose levels, but a balancing act. CF care therefore needs be individualised and constantly evolving to meet these changes in clinical status.

The concepts which emerged from this study are similar to those seen in other qualitative evidence syntheses concerned with the experiences of managing multi-morbid conditions (Liddy et al. 2014, White et al. 2016, Cheng et al. 2018). These included maintaining a normal life, keeping a positive mental attitude, adapting to change, hierarchical ordering between chronic diseases and coping in a social context (Cheng et al. 2018). In this study, CF was often seen as the most important illness because it is life-limiting but having CFD further impacts survival (Lewis et al. 2015). This is not unique to the management of CF and CFD as people often have to prioritise conditions or personal values to manage multi-morbid conditions (Liddy et al. 2014, White et al. 2016, Cheng et al. 2018). Trying to retain a positive mental attitude and developing skills and coping mechanisms so individuals can adapt to change are important factors in trying to maintain a 'normal life' when managing multi-morbid conditions (Liddy et al. 2014, Cheng et al. 2018). Despite the differences in signs, symptoms and effects of different multi-morbid conditions the experiences of people with these conditions are comparable.

Some people with multi-morbid conditions reported being overwhelmed at times but many more appeared to be resilient and determined to persevere despite challenges (Liddy et al. 2014). In the CFD populations within this study there was not a sense of being overwhelmed, CFD was seen as more of an inconvenience and nuisance. It led to disruptions in daily life. It challenged identity by bringing illness into the foreground because treatment needed to be conducted outside the home and could no longer be hidden. Experience developed while living with one chronic condition and familiarity

with healthcare systems can help the management of subsequent illness (White et al. 2016). The majority of people with CF have attended hospital and clinics from a very early age. Through this they have developed knowledge of their condition, experience of managing complex treatment regimens and become familiar with healthcare systems. This would have facilitated the adjustment needed to accommodate CFD into their lives, with more successful management seen when CFD was managed as part of CF and incorporated into pre-existing routines.

Only one systematic review into the experiences of living with CF has been conducted; this involves children rather than adults (Jamieson et al. 2014). However, there are similarities in the experiences observed which include: gaining resilience (regaining control, redefining normality), lifestyle restrictions, resentment of chronic treatment, temporal limitations (taking risks, valuing time), emotional vulnerability and transplant expectations and uncertainty (confirmation of disease severity). Children with CF become adults. What informs their self-identity experiences and beliefs will be with them for life, and are therefore important foundations of who they are and what CF means to them.

### 3.3.1 Strengths and limitations

This meta-ethnography demonstrates that there is very limited qualitative research exploring the experiences, particularly with regards to feelings and emotions, that adolescents and adults encounter in living with and managing CFD. However, it increases the evidence base and enhances the strength of its findings (Doyle 2003) by including unpublished work (a research thesis) which was previously only available as an abstract for the 2012 European CF Conference (Richmond 2012). This qualitative research increases knowledge of patients' experiences of CFD and this will contribute to identifying changes needed in patient care.

Meta-ethnography brings together studies to create a new interpretation (Noblit & Hare 1988). However, the interpretative nature of meta-ethnography recognises that there is variability in interpretations between researchers due to differences in personal experiences and knowledge. My dietetic professional background, training

and experience therefore influenced my interpretation of the findings. The wider long-term condition perspectives of other members of the research team contributed to exploration and challenges in making interpretations. Challenging personal interpretations are vital to ensure rigor in qualitative research (Toye et al. 2014). Therefore, the use of more than one reviewer within this study contributed to increasing rigor. The key findings were discussed with the study project management group, which consisted of lay and professional representation and this therefore contributed to enhancing the credibility of the findings within this part of the study.

The author of this meta-ethnography was the first author of one of the included papers (Collins & Reynolds 2008). Meta-ethnography does not involve the re-analysis of data; it uses the author's interpretations of their data (concepts) as its data. The concepts from Collins and Reynolds (2008) were not treated any differently from those of the other included studies, therefore only the published work within that paper was subject to analysis. Other meta-ethnography studies have been published where one of the authors contributed to a paper within the synthesis, without this being acknowledged (Britten et al. 2002, Pound et al. 2005, Hughes & Noblit 2017). Being open about this and the acknowledgement of treating all concepts equally contributed to transparency within this study.

Quality appraisal has received criticism in meta-ethnography (Atkins et al. 2008, Toye et al. 2013, Toye et al. 2014). This study asked two screening questions, prior to quality appraisal, these were: does the study report results from qualitative research and did it involve both qualitative methods of data collection and analysis? and is this research applicable to the synthesis topic (Pound et al. 2005). These questions were more helpful than CASP screening, which was time consuming. If these two criteria were not met, the studies were excluded. CASP focussed on methodological flaws in qualitative research and none of the included studies had major methodological flaws. Using CASP therefore didn't contribute any more than just using the two screening questions. The two mixed-methods studies included less supportive qualitative data, but still added to the experiences (Dashiff et al. 2013, Millington et al. 2014). Each study therefore contributed to this synthesis.

Meta-ethnography reporting guidelines were published on 15th January 2019 (France et al. 2019) this was after this meta-ethnography was completed. The guidelines consist of 19 reporting criteria. Each of these reporting criteria have been subsequently compared in relation to this study (appendix 6). All of these reporting criteria were met within the study, the criteria concerning how the studies were related to each other (number 11) was brief and could have benefitted from a bit more detail.

### 3.3.2 Implications for practice and further research

In agreement with recommendations from the majority of studies that formed this meta-ethnography, CFD management needs to be comprehensive, integrated into routine care, and adaptable to consider changes in health status (Collins & Reynolds 2008, Tierney et al. 2008, Dashiff et al. 2013, Millington et al. 2014, Sylvain et al. 2016). CFD should be regarded as a natural progression of CF, these two conditions are so closely intertwined. Normalising CFD management as part of routine CF care will help achieve a sense of balance because all treatments, therapies and self-care strategies are integral to each other.

As discussed in chapter one, this meta-ethnography was the driving force to reconsider the name cystic fibrosis related diabetes. On reflection of the findings it was felt that the term 'related' does not infer a direct causal relationship but more of an association. The name cystic fibrosis diabetes, which is in line with type 1 diabetes or type 2 diabetes, is considered a more appropriate term and was adopted as a preferential term in this thesis. Adopting the name CFD in clinical practice would encourage people with CFD and their healthcare professionals to see the condition as a combined entity, as per figure 15, and influence approaches to care and management.

Adjustments to the new health status needs to be recognised following new diagnosis and identity need to be considered by healthcare professionals as illness perception can change. People with CFD need help preparing for how treatment is managed outside the home to reduce feelings of self-consciousness about the visibility of diabetes. All people with CFD should therefore receive appropriate wellbeing informed

care, from healthcare professionals as a continual part of their care. This should include assessing for difficulties in adapting to diabetes at diagnosis and changes in health perceptions that may occur due to fluctuations in clinical status.

CFD is the most common co-morbidity affecting people with CF, however despite earlier screening and treatment survival in people with CFD remains reduced. Further research is needed to explore CFD self-management experiences and requirements, and to how to best deliver appropriate support and education to these people. This research should adopt a qualitive approach to obtain detailed accounts which will reflect individual experiences and the diversity seen in CF populations. Findings from this meta-ethnography which need further investigation include: exploring the thoughts and feelings that people with CFD hold about the perceived relationship between CF and CFD; reviewing self-management behaviours - this should include looking into barriers, facilitators and successors of CFD self-management; identifying what individuals require to manage their CFD on a day to day basis - this should consider support mechanisms, resources and knowledge. This research will help improve CFD management and knowledge and optimise glycaemic control as a significant contributory factor to survival (Adler et al. 2011).

## 3.4 Conclusions

Striking the balance between CF and CFD highlights that individual perception of CFD directly affects its management. The close link between nutritional status, lung function and glycaemia is a fundamental part of management and directly influences survival. CF and CFD are intertwined; there is a direct relationship between them. CFD therefore should be regarded as part of CF and education, psychosocial support and management should be tailored around this, as part of routine care, rather than separating out CFD management. Education should not just focus on people with CFD but must also include healthcare professionals and families/carers. Further research is needed to explore self-management support experiences and requirements. It is also time to reconsider the name cystic fibrosis related diabetes and adopt the name cystic

fibrosis diabetes which acknowledges that diabetes is not associated with but caused by CF.

## 3.5 Research Outputs

Striking the balance between cystic fibrosis and Cystic Fibrosis Diabetes (CFD) - a meta-ethnography. Oral and poster presentation at CATO Research Symposium, Hammersmith Hospital (27th June 2018).

Collins, S., Jones, A., Woodward, S. & Sturt, J. (2018a) P160 Striking the balance between cystic fibrosis and Cystic Fibrosis Diabetes (CFD) - a meta-ethnography. *Journal of Cystic Fibrosis***17**, S104. Poster presentation at European CF Conference, 6-9th June 2018.

Experiences of living with and managing CFD. Oral presentation at adult and paediatric CF research away day, Royal Brompton & Harefield NHS Trust- (27th March 2018).

Striking the balance between cystic fibrosis and Cystic Fibrosis Diabetes (CFD) - a meta-ethnography. Poster presentation at Royal Brompton & Harefield NHS Trust's Nursing and Allied Health Professional Research Day, 30th October 2017. Awarded best respiratory poster award.

PhD project management group meeting 1, 20th April 2017- overview of PhD and presentation of meta-ethnography.

# 4 What are the self-management experiences of people with cystic fibrosis diabetes?

## 4.1 Methods

### 4.1.1 Research design

A qualitative single occasion interview design, guided by the principles of IPA (Smith et al. 2009) was used to answer the question "what are the self-management experiences of people with CFD?" IPA is concerned with understanding and exploring the lived experience of a particular phenomenon; specifically, the meaning people attach to their experience (Smith 2004). The theoretical underpinnings of IPA (as discussed in chapter two) are idiography (individual perspective), phenomenology (person's direct experience) and hermeneutics (interpretation) (Smith et al. 1999, Smith 2004, Smith et al. 2009).

With the aims being:

- To confirm the findings from the meta-ethnography prospectively and to identify any additional concepts
- To establish what are the barriers, facilitators and successes that people with CF experience in managing their CFD
- To identify knowledge and skills people with CFD require and use for self-management of CFD and how they might be incorporated into a programme
- To help identify content for a CFD self-management education programme

All members of the research team: SC, JS, SW and AJ contributed to the IPA study. SC led the IPA study by collecting and analysing the data. Five of the transcripts were independently reviewed by members of the research team (transcript 1- JS, SW, transcript 3- SW, JS, AJ, transcript 5 – SW, AJ, transcript 6 – SW, JS and transcript 7 -AJ) to explore and validate identified themes and ensure reliability. These roles are acknowledged throughout this chapter with the individual's initials. The key findings were also discussed with the study project management group, which consisted of lay and professional representation.

### 4.1.2 Research setting

All participants received their care at a Regional Adult CF centre. All interviews took place in a private room within the research facility at this CF centre.

### 4.1.3 Participants

Participants were recruited from the CF centre's patient database. IPA works with homogenous samples therefore, the participants were purposely sampled to meet the inclusion criteria of having CFD, being over 16 years of age and taking insulin (table 20).

**Table 20 Inclusion and exclusion criteria**

| Inclusion criteria | Exclusion criteria |
|---|---|
| • CFD -treated by insulin<br>• >16 years old | • Treated by oral hypoglycaemic agents (OHA) alone (currently <2% of centre's CFD adults take OHA)<br>• Inability to participate in an interview due to instability or severity of underlying cystic fibrosis lung disease<br>• Unable to understand/read English |

Forty-four letters were sent out, to people identified as meeting the inclusion criteria, inviting them to take part in the study. Eight people responded to the letters and agreed to participate in this study. The remaining 36 people did not respond. All participants were known to the researcher in a clinical capacity, therefore so as not to coerce people into participating, follow up contact was not made. Letters of invitation were sent out in stages until the recommended number of participants were recruited.

The idiographic nature of IPA leads to the recommendation of a smaller sample size; the main concern of IPA is with the detailed account of individual experiences. Guidelines for what constitutes a reasonable size for when using IPA have been suggested as three to six participants at undergraduate or master's level and between four to ten for professional doctorates, however sample size is contextual and should be considered on a study-by-study basis (Smith et al. 2009). In his review evaluating the contribution of IPA research Smith (2011) refers to larger IPA studies as having

more than eight participants (Smith 2011). However there is no correct answer to what the sample size should be (Smith et al. 2009). There is the need for each individual perspective to understand how the phenomenon is experienced but cross-case analysis is needed (unless it is a case study) to be able to move closer to the core of the phenomenon. Practical considerations such as time frames, the research question and participant availability also need to be considered. Eight people with CFD were interviewed between July and October 2017, this is a typical sample size seen in IPA research (Smith 2011). Participants received a £20 'thankyou' voucher for taking part in the study, and if required travel expenses were reimbursed.

### 4.1.4 Ethical considerations

Prior to commencing the study approval was received from: London-South East Research Ethics committee- 17/LO/0377 (appendix 7), NHS Health Research Authority (appendix 8) and Royal Brompton and Harefield NHS Foundation Trust (appendix 9). Informed written consent, including agreement for digital audio-recording, was obtained prior to starting the interviews. A copy of the participant's consent form was stored in their electronic patient record. A private transcription service was used for transcription. To ensure confidentiality all recorded interviews were uploaded via an encrypted server to the transcription company. A confidentiality agreement was agreed with the transcription agency (appendix 10). All names, places and other identifiable details were then removed from the transcripts prior to analysis.

### 4.1.5 Data collection

Semi-structured interviews were used; the interview topic guide focussed on the experience of living with and self-managing CFD. Existing literature (Collins & Reynolds 2008, Segal 2008, George et al. 2010, Withers 2012, Dashiff et al. 2013, Millington et al. 2014, Wilkinson et al. 2014) and the findings from stage 1a (chapter three) provided key themes and prompts for the interview topic guide which focussed on: living with CF, the diagnosis of CFD, managing CFD, the impact of CFD and self-management (see appendix 11). From existing literature key areas concerning self-management in people with CF and diabetes were considered when constructing the interview topic guide. The perception of the barriers and facilitators informing CF self-management decisions

in older adolescents and adults with CF (George et al. 2010) such as the influence of work and social demands and the role of CF clinic were considered an important part in informing CF self-management decisions. In adults with type 1 and type 2 diabetes key factors influencing the ability to self-manage included five key areas namely: communication, education, personal factors (beliefs, physical, practical, psychological), provider issues and support (Wilkinson et al. 2014). Studies by Collins & Reynolds (2008) and Millington et al. (2014), although included in the meta-ethnography, provided additional information to inform the interview topics guide and its prompts as their papers included examples of their interview questions used within their studies. Questions and prompts in the interview topic guide were designed to reflect the five keys themes identified in the meta-ethnography: a need to develop a perspective on CFD, struggling with identity, a representation of morbidity and mortality, in control or being controlled and need for knowledge and support (appendix 12).

The interview questions were open-ended and designed to be used as prompts, this gave the interview schedule greater flexibility. Participants were encouraged to tell their experiences in their own words; the questions were intended to facilitate this process of gathering rich, in-depth data rather than narrowing it to predetermined categories. Interviews were digitally recorded and transcribed verbatim.

### 4.1.6 Data analysis

Interview transcripts were analysed using the process of IPA, which consists of several stages. These stages are summarised in table 21 (Smith et al. 2009). Examples of the data analysis process are given in appendices 13-18. The process of data analysis is systematic, but it is designed to be flexible and not prescriptive. Researchers need to become close to the data with the aim of trying to step into the participant's shoes as far as possible (Pietkiewicz & Smith 2014). It is therefore important to ensure that the participant is the focus of analysis as the process of entering their world begins. Exploratory coding has three main areas of interpretation: descriptive, linguistic and conceptual (Smith et al. 2009). Focus should be on that particular moment, staying close to the meaning, paying attention to what is going on, what language is being

used and what does the data actually mean. These three steps of exploratory coding are unique to IPA. Table 22 highlights the focus and practical application of exploratory coding. Data analysis consists of word-for-word, line-by-line review. The focus of data analysis is to remain on the sense-making experience from the individual to the shared and from description to interpretation (Smith et al. 2009).

**Table 21 Stages of IPA**

| Step | | Process | Example |
|---|---|---|---|
| **Step 1** | **Reading and re-reading** | Familiarise and stay close to the data | |
| **Step 2** | **Initial noting** | Word-for-word, line-by-line review<br>Exploratory coding- descriptive, linguistic, conceptual | Appendix 13 |
| **Step 3** | **Developing emergent themes** | Aim to map interrelationships, patterns and connections between explanatory notes<br>Transform initial notes into more specific themes and phrases | Appendix 14 |
| **Step 4** | **Searching for connections across emergent themes** | Establish connections between themes<br>Organise into clusters of related themes | Appendix 15<br>Appendix 16 |
| **Step 5** | **Moving to the next case** | Move to the next transcript<br>Repeat the process | |
| **Step 6** | **Looking for patterns across cases** | Compare and contrast clusters across participants<br>Look for connections between each participant | Appendix 17 |
| **Step 7** | **Development of super-ordinate themes** | Clusters may be merged together to form super-ordinate themes | Appendix 18 |

**Table 22 Focus and practical application of exploratory coding**

| Exploratory coding | Focus | Practical application |
|---|---|---|
| • **DESCRIPTIVE** | describing the content of what is said | Subject of talk within transcript<br>What jumps out at you? |
| • **LINGUISTIC** | exploring specific use of language | Look for unusual, imaginative or quirky words or phrases<br>Use of metaphors |
| • **CONCEPTUAL** | engaging more at an interrogative and conceptual level | What does the data actually mean?<br>Challenge and question underlying meaning |

First, ensuring an idiographic approach, each transcript was analysed individually before moving onto the next (SC). Each transcript was read multiple times making descriptive, linguistic and conceptual notes in the right-hand margin of the transcript (appendix 13). Once this was completed for the whole transcript, I returned to the beginning of transcript and began to develop the exploratory comments into a set of emergent themes that highlighted key features of experience developed from the participant's account. A Microsoft Excel workbook was created, to assist with data analysis, for each participant. In this, emergent themes were recorded with illustrative quotes from the participant to ensure that they reflected the participant's meaning (appendix 14). The emergent themes were then merged together to reflect their shared meanings (appendix 15). This was carried out by visually exploring all the emergent themes which were recorded in the Microsoft Excel workbook. These themes were then further clustered together and given a label which reflected the shared meaning or the wider concept of the theme (appendix 16). The next case was then moved onto and the process repeated. Once all the transcripts had been individually analysed a process of cross-case analysis was undertaken. Table of themes for each participant were compared, with close attention paid to instances of divergence and convergence (appendix 17). The themes were grouped, according to similarities, into subordinate themes and these were subsequently organised into

superordinate themes which focussed on a central concept (appendix 18). Data analysis continued throughout the process of writing up, where closeness to the data remained.

### 4.1.7 Quality in qualitative research

Yardley's work (2000) has been recommended to provide a framework to evaluate and demonstrate the quality of IPA research (Smith et al. 2009, Hefferon & Gil-Rodriguez 2011). It provides a flexible approach, with many opportunities, to assist IPA researchers to demonstrate quality in their work (Smith et al. 2009). The four key dimensions for quality in qualitative research namely: sensitivity to context; commitment and rigour; transparency and coherence; and impact and importance were considered throughout this study (Yardley 2000). Sensitivity to context was demonstrated through the analysis by giving participants a voice through verbatim quotes; this also allows the reader to trace back the interpretations. Commitment to rigour was demonstrated through the detailed commitment to the idiographic and in-depth nature of IPA, by following guidelines for representing the identified themes in an appropriate number of participants (Smith 2011) and by involving more than the researcher in the data analysis process. I conducted the primary data analysis, and the supervisory team (JS, SW, AJ) reviewed five transcripts to explore emergent themes and interpretations at the end of each individual case and on completion of cross case analysis. Interpretations and emerging themes were challenged to ensure the analysis was thorough and grounded within the data. Each step of the research process is clearly outlined in the data analysis section to aid transparency.

#### 4.1.7.1 Reflexivity

In IPA it is important to consider how the researcher's experience, role and knowledge influenced data collection, analysis and interpretation. I acknowledge that my experiences of working with people with CFD for over 20 years has informed the research process and may have influenced how I have collected and analysed the data. I also knew some of the participants in the study reasonably well, and had my own understanding of them, their life and disease management. Throughout the interviews I tried to put these assumptions aside and sought clarity of individual experiences by

asking prompts rather than being guided by my assumptions or prior knowledge. As IPA involves double hermeneutics whereby the researcher interprets the participant's interpretations of their experience it is important that the researcher is aware of prior assumptions and knowledge that might influence their interpretations. A reflective journal was kept throughout the research process in order to raise my awareness of any difficulties I encountered during data collection and analysis and to acknowledge any pre-conceived thoughts or feelings that challenged me. This awareness helped me to be more open-minded and less judgemental. I was able to personally reflect upon any difficulties encountered and how they impacted upon me. Where I felt that I needed further assistance I sought help through supervision or from peer support at a regional IPA group.

My role and how participants within this study would have previously known me is an important consideration. I was a clinician who was part of the CF team delivering clinical care to these individuals, but for the purpose of this study I am a researcher. It was important to remain mindful that some participants may struggle with this. This could lead them to withhold information as they may be concerned about the impact of what they say upon their clinical care or they may respond in more favourable ways. To help with this my role in this study was discussed with participants prior to the interview.

## 4.2 Results

Eight adults (4 female), aged between 22-48 years (median 31.5 years), participated in the interviews. Seven were on basal-bolus insulin regimens and one was just on basal insulin. Participants HbA1c ranged from 5.3-10%/ 35-86mmol/mol (median 7.6%/ 59.5 mmol/mol); BMI ranged from 17.7-23.9 kg/$m^2$ (median 21.6kg/$m^2$) and lung function ($FEV_1$) ranged from 33-81% (median 50.5%). The interviews for six of the participants coincided with visits to the hospital for clinic appointments. The other two participants chose to attend for interviews at a different time convenient to them. The interviews lasted between 49 and 82 minutes (median 57 minutes). All participant's names referred to in the data analysis are pseudonyms.

The findings are presented as superordinate themes, subordinate themes and participants' quotes. The superordinate and subordinate themes identified from the data analysis are listed in table 23.

**Table 23 Superordinate and subordinate themes**

| Superordinate theme | Subordinate theme |
|---|---|
| **Forming a relationship with CFD** | CFD- impact, constancy and exposure<br>The conflicts, complexities and unpredictability of CF and CFD<br>Facing morbidity |
| **Balancing the CFD self-management triad** | Insulin- understanding and uncertainty<br>Blood glucose monitoring – comprehension, confusion and compromises<br>Managing nutrition- the challenges and frustrations |
| **The unmet need for information and support** | The need to feel informed, skilled and supported<br>Creating systems to deliver information and support |

## 4.2.1 Forming a relationship with CFD

This superordinate theme explored the relationship that participants developed with their CFD from diagnosis through to incorporation into daily life.

### 4.2.1.1 CFD- impact, constancy and exposure

The diagnosis of CFD was met with many emotions and feeling, most of which were initially challenging to deal with, but diminished with time.

> *Erm well you're not delighted, obviously. I mean you'd be pretty upset, pretty upset that you'd gotta do it. But then, as I say, I think because I was so unwell it was – and then between that and the overnight feeding you saw a gradual improvement and you could see it made sense to do it. (Tom)*

> *Erm, at the time, I think I, I was a bit upset with it but I think I've been like that kind of every time you get a new diagnosis. You know, I'm upset for about a day and then it's like, 'Right, what can we do?' and you just kind of get used. (Ana)*

For Paul it wasn't the diabetes itself that was "hard" for him it was the development of "something else to keep an eye on and something else takes concern," that "extra thing to think about."

The significance of diabetes became more apparent and significant with the realisation that it was not going to go away especially for some, as it was a sign that their health was starting to decline.

> *Erm, so at the time, it was like, 'Okay'. I hadn't understood really what that meant. Erm, to me, it was just – Oh, just another treatment but actually, it was – you have diabetes. It's not going to change. It's not reversible and not only that, you need to take insulin forever. (Brian)*

> *I mean it's an indication that your body is... it's sort of chipping away or like there's a deterioration that erm, is, is causing permanent – this is, you know, now permanent and erm... and that's – you know, that, that idea that you're on a downward slope, rather than up and down is er, is quite heavy. (Ben)*

Having a pre-existing routine to manage CF helped with the management of CFD by incorporating the treatments together; this lessened the impact of CFD.

> *so I'll go and eat, when I'm sitting down for a meal making sure that I can have my insulin beside my creon pot because I always go for the creon pot because I need to have that so there's the insulin beside it which makes sense. Er, keep things together, trying to build patterns and routines as much as possible. (Paul)*

> *on a day-to-day level, it's just kind of – it's almost automatic. You know, it's just like taking those tablets in the morning. I just get up and do it and you just get on with your day kind of thing. You just kind of – you go into just automatic robot. You don't even notice what you're doing really. (Ana)*

"Just got on with it," "just took it in my stride" and "adjust to new normal" are examples of positive attitudes and coping strategies which aided the participants ability to manage the addition of CFD.

For Mary, CFD was more in control of her than she was of it. Despite her engaging with trying to identify her problems with blood glucose management she showed feelings of despondency and frustration. There was a sense that Mary felt alone and was trying to manage this on her own. Reminders of the damaging effects of CFD further added to the spiral of loss of control for her, rather than helping her.

*Because I just can't get to grips. You know, you, your, erm, sugar levels even when I think that I'm trying my best just don't seem to work out. You know, I, I had, er, an implant thing on my arm once for three days, I think Dr XXX done that. I wrote down everything I ate, this, that and the other and they were still coming out high and I just think, 'Where, where am I going wrong? What am I doing wrong?' And it's, it's running my life because I can't get it under control. And then the doctors say it's like a vicious circle. Sugar in your blood, then the bugs eat off your blood and then you get chest infection and it's like a vicious circle so yeah, that's how I feel it controls.*

The constancy and invasiveness of CFD had a significant impact upon life and was a barrier to successful management. Ana described the analogy of having CFD as "it's like a pet in a way," she related this to something that also needed constant monitoring and feeding regularly to "keep it happy." Planning and constant attention around the management of CFD in daily life were seen as an inconvenience and annoying.

*The injection and it – I think it's the invasiveness of the treatments; so the invasiveness of doing all the BMS erm, and then they're minor, they're minor compared to other things but actually, erm, having to inject and do things like that, I just, I just find it irritating [laughter]. (Brian)*

*Erm, it can, can be a bit of a pain because […] I have to plan and organise, you know, my meals, what I'm going to have, erm, what snacks I can, I need to think ahead constantly. I'm already doing that with my CF and I have to do that again for my diabetes. (Karen)*

Difficulties coping with the constancy of CFD were not just observed in the initial period of learning to live with CFD. Brian has had CFD for more than 20 years and the constancy required to manage CFD was still problematic for him. He demonstrated honesty by confessing he wasn't managing his CFD. A fact that he appeared to be hiding behind.

*..but in my heart, I'd know I'm not really managing it but actually, I am, I am, I am managing better erm, but it's, it's the consistency with me and this is the thing. (Brian)*

For Mary, who found CFD management exceptionally challenging, the presence of CFD disrupted her life. She was angry and frustrated. The constancy of CFD management for her wasn't the doing but the thinking about what the not doing meant. Mary was insightful that her CFD needed more consideration, but she described no actions indicating that she was responding to this insight.

> *My CF was in a routine. My diabetes makes my life not in a routine because it's constantly there. It's 'cause you're constant, even though I don't do it I'm, I constant, I do think about it and everything and the repercussions and everything but it's something that I think needs to be more looked after than what my CF is, do you know what I mean? (Mary)*

Many treatments for the management of CF are conducted privately at home; these therefore do not attract the attention from others. Unlike treatments required for CFD such as injecting insulin and the monitoring of blood glucose levels which are very visible to others. This visibility generated a sense of unease and an awareness of consideration needed towards other people.

> *it's very inconvenient to be checking all the time you know when you're out and about and you have to get your finger pricker out and yeah it's you can't really do it well I don't feel comfortable doing it in front of strangers erm you know it's the same with insulin if you're out and about some people will just do it in front of people I'm much less comfortable doing that I'd rather take myself off to a loo or something private and if that's not accessible then I will wait until I can get to a place where I can do it privately which isn't great because that's when erm if I've eaten then I have to wait before I do my insulin and stuff like that. (Jane)*

> *I find it really difficult doing injections, er, when out with other people. Erm, it's finding – finding the space to do it, finding the right sort of appropriateness. (Paul)*

Brian displayed a sense of shame and stupidity that he could not possibly admit after many years with CFD that he needed some support for him be able to manage his CFD.

> *I suppose the other bit – and this is going to sound really weird – you know, I'm – but I've had it [CFD] for over half my life – longer and I sort of feel it would be a bit strange, for all intents and purposes, for me to admit that I need a bit of help or that I could do with someone just sitting down with me and sort of*

*pressing reset. It, it sounds weird.[…] In my mind, I'm thinking, 'You kind of can't – you can't do that. Don't, don't be stupid' erm, but yeah, be-, being honest, that's...*

Mary described CFD as being a "pain in the arse and brain damage." She was being displayed as a failure as she could not control or cope with CFD. However, on a positive note, following psychological support Mary experienced a huge sense of relief and realisation she was not the only person with CFD to feel this way.

*Because it does my head in. It really does do my head in. Yeah. I've seen a psychologist twice in here about it […]. Erm, I said to her, 'I feel like it controls me', and, and do you know what she said, you know, which really helped. She went, 'You're not the only person that feels like that', and I honestly thought I was. And I do, and I did, that's the truth. I thought I was on my own. I, I was the only one that couldn't cope but she said, 'No, you're not'. She said, 'Other people, you know, are out there as well', she said, 'And they're, they're not dealing with it', which made me feel a bit better because I, I did, you know, it's the only thing that I struggle with.*

#### 4.2.1.2 The conflicts, complexities and unpredictability of CF and CFD

One of the major challenges in the management of CFD was how the relationship between CF and CFD was viewed and managed, however this was not straightforward. There was a hierarchical comparison between CF and CFD: the impression that CF was developed first, was more significant and the effects of not doing treatment for this were more serious and instantaneous when compared to that of CFD. CF impacted more on "physical" health than CFD, this would likely be due to the associated effects of CF on respiratory function.

*I suppose when it comes to kind of the, the crude way of looking at it, it's the consequences of me not doing something for my – for the CF side of things are more immediate than the diabetes side. I know it's the wrong way but it's just how I can sort of rationalise maybe some of my wrong choices. (Brian)*

*[…] if I skip NovoRapid with a meal, I won't, I won't feel it. I mean if I were to get up and leave the house without taking m-, my Salbutamol, or Seretide, or DNase, or... or nebulised antibiotic, I'd probably – I'd feel significantly worse. (Ben)*

*Oh my god. It's nothing' [laughter]. It's [CFD] nothing in the grand scheme of things compared to, I think, again like, you know, monitoring my lungs. (Ana)*

There was ambiguity in some participants if CFD was part of CF or something different, with links between the two conditions mainly concerning medical treatment and dietary management.

> *[…] I see them linked and interacting, erm, because of, particularly because of medical, medicine interactions, erm, and dietary. (Paul)*
>
> *I probably see it as being part of my CF. But I use this 'probably' to justify getting away with eating stuff I shouldn't because it doesn't feel like proper diabetes in inverted commas. (Tom)*

Mary and Brian, who have tended to regard their CFD as a separate condition experienced more issues with trying to achieve a sense of control and balance.

> *Well because they are separate. One's CF, one's diabetes, isn't it? And I can see that, the thing between the two of them like I said with that vicious cycle but they are two different things, aren't they. Yeah. (Mary)*

Brian's separation for the management of his CFD goes beyond just managing it. It was part of his partial acceptance as he believed CFD was "something that's been done" to him as he developed it related to steroid therapy. He says "*I've kind of boxed diabetes off as a separate entity and I've, and I've always kind of treated as a, as a – to punish it, in my mind."* However, Brian did reflect that his strategy to management was not the best and his advice to others would be to treat them as one condition.

> *'It's part of CF and this is how you're – how to manage it in conjunction with the CF. Don't do what I did and treat them as two separate entities. Sort of entwine them as one (which I suppose they are) erm, and have a treatment plan and a regime that complements both'. (Brian)*

How the CFD service was delivered further impacted upon CFD management. The centres specialist CFD clinic was held on a different day to general CF clinics, with CFD specialists, thus by dividing up the service healthcare professionals are influencing how people with CFD construct their understanding of it. The provision of service in this manner is itself contributing to a barrier in management.

*It does feel like it's separate. And I don't know as well if – it might be different if you saw someone in a CF clinic for CF related diabetes at the same time. So psychologically you're going to a different clinic and that sort of in your mind separates it as well. (Tom)*

*So, if I was having an issue with the diabetes yeah okay that needs fixing but again its straight back onto the chest so I think erm its almost that even in clinic the two are separated very much. (Jane)*

Participants demonstrated knowledge that there was a relationship between high blood glucose and respiratory pathogens. The direct link between high blood glucose and the harmful consequences it has on CF respiratory symptoms was a significant factor in promoting management.

*the only thing that directly affects me is the, well the side effects of the high sugars onto the chest. (Karen)*

*I do accept that when my blood sugars are, are, you know, continually high that when I've got bugs in my chest, they thrive off the glucose and high blood sugars. So that's always in the back of my mind to try and stay on top of. (Brian)*

*I can see what the doctors are saying. I see the vicious cycle of the sugar in your blood, then the bugs feed off your blood where the sugar is and then you get obviously the bacteria and the phlegm in your chest, which can cause a chest in, I can, I can see that. In paper and in my mind, I can see that cycle. (Mary)*

Living with CF was viewed as being "unpredictable." This contributed to difficulties in managing CFD. CF directly impacted the management of CFD and changes in treatments or respiratory symptoms contributed to the unpredictability and warranted more attention; that was something else participants reported they needed to think about.

*suppose yeah, I mean day-to-day, it's, it's – tends to be – I'm pretty routine about it but if – I suppose yeah, if, if I'm feeling worse or I've had a change in medication, then it's a fear that that – that's about – it's adding – that my high blood sugars are adding to my poor situation; that that, that I'll check it more to make – just make sure that that's under control and that's not an a-, - an additional factor. (Ben)*

*I think it, erm, I think maybe once when I had an infection it was a bit less, well it was, it wasn't as easy to control and it maybe required more insulin or my, my*

*sugar was low even though I was eating loads and insulin, like doing my normal dose of insulin. Erm, so I think it [CF] can sometimes make my sugars slightly sporadic and* unpredictable but that's more when I'm having IVs I think. (Karen)

*That's what I find quite difficult is that it can be quite erratic the erm yeah to if I can eat, if I eat the same things, do the same things, have very similar days, my insulin requirements can be quite different and I've always put that down to maybe something different is happening with my chest and that's causing you know slightly different, different readings so that's why it can be quite difficult to keep on top of as well erm because it isn't just as straight forward as have the same doses all the time so yeah. (*Jane)

The complexities of trying to manage CFD was highlighted by Jane. She had identified a relationship between CF and CFD by suggesting "the worse my chest is the worse my diabetes is and the worse my diabetes is the worse my chest is" but she acknowledged "trying to keep them both under control is incredibly difficult." However, Jane recognised this relationship did not always exist, there was an element of unpredictability and therefore multiple layers of complexity that were not always easy to understand.

*the two are very much directly related and just because just if my CF is good doesn't mean that my CF is, if my chest is good doesn't mean my diabetes will be good though if that makes sense? And if my diabetes is good it doesn't mean my CF will be good it might just be easier to manage but I can have one bad and one good. Does that make any sense? (Jane)*

Participants who received steroid treatments as part of their CF management experienced elevations in their blood glucose levels that proved very challenging and aggravating. A new sense of loss of control was seen, accompanied by a sense of looking for a new understanding.

*it's taking that prednisolone it's whacked up to 40mg and you just see your blood sugars shoot through the roof. Erm, and yeah I think the first time I remember seeing that particularly at the time I was taking 10/12 units of NovoRapid with a meal time and over that inpatient stay that was more than doubled just to try to get on top of my blood sugars. (Paul)*

*steroids you know you're doing that's actually something beneficial for your CF but it then throws you, all your diabetes into yeah it goes haywire totally and it,*

*that's always difficult to then especially with a constantly changing dose of steroids to keep on top of that yeah as well as all the other things so. (Jane)*

*Erm, I just – yeah, it was – the sugar just seemed uncontrollable erm, and... yeah, I just didn't really know – th-, that – I think, I think part of the reason I found it difficult was because I had come from such a stable erm, diabetic control and this was kind of catapulted in – into a complete rollercoaster, you know, and I was getting extreme highs of like, you know, 18-20 erm, and it was – you know, I was on really high steroids. (Ana)*

*I've been on a high dose of steroids and had like a nasty chest infection as well and it's – there just doesn't seem to be enough insulin that you can put in your body to – for it to get down to. (Ben)*

There was a sense that healthcare professions had not prepared participants for the consequences of steroid treatment or provided sufficient support and reassurance during the course of treatment. Participants experienced fear and uncertainty over what to do. There were both emotional and physical barriers to increasing insulin doses, which created a real fear of the unknown which for some could be drastic.

*for me, in a way erm, I was almost psychologically afraid to give that much insulin to myself because I was like, 'I'm definitely going to low. That's going to kill me. I'm going to go low and go unconscious.' (Ana)*

Paul was trying to balance a conflict between being told by the team *"not to worry about" the* sugars as they were *"just going to deal with the chest because the sugars will sort themselves out [...]"* This response challenged Paul and he didn't want to be defeated as he was aware of the *"long term implications"* of high blood glucose. The experience was like a battle to Paul, with opposing sides and significant consequences.

Brian had an extremely negative relationship with the use of steroids. He resents steroids and blames them for causing his diabetes, a problem that he has never felt that he has fully come to terms with because he felt it was inflicted upon him during his adolescence.

*in my mind, I had a chest infection, what I was being given to help with the chest infection, as a side effect, has caused me to become diabetic and that's a real struggle. I was very angry about that – very angry for quite some time that – erm, the speed at which it all just happened.*

#### 4.2.1.3 Facing mortality

Thoughts for the future were predominated with CF rather than CFD. Participants were very aware of the limiting effects CF will have upon their lives. For this reason, CF management was often prioritised over CFD.

> *I know that I'm going to die from CF and not from diabetes [laughs]. (Mary)*
>
> *No, 'cause the lungs will kill me first [laughs] so if I'm honest. (Karen)*
>
> *CF is the more here and now erm, and that, for me, is what's going to, in my mind, trip me up first. (Brian)*

Participants were aware that they were at risk of diabetes related complications, especially eye damage but they didn't ruminate on these, they just put their thoughts concerning this in the background.

> *the idea of my eyesight deteriorating as a result of that is quite a concern to me. I ha-, - I mean, luckily er, so far, they've told me that it hasn't affected my eyesight at all but like the i-, - the idea that it would is – as well as it affecting my chest is something that I don't want – I don't, I don't want to have. (Ben)*
>
> *I'm aware of, you know, the possibilities with – when you inject and when – in terms of your blood sugar levels and the tedious effect it can have on your sight and whatnot. But erm – so yeah you're obviously aware of that. (Brian)*
>
> *when you go and get the annual eye test sort of okay, how are things, is this realistic you know that, er, I'm going to end up losing part of my sight, is that something that's conscious but it's something that you know you get looked at every year but you just sort of know that that's there. (Paul)*

### 4.2.2 Balancing the CFD self-management triad

Blood glucose, insulin and nutrition were identified as the most influential components in the self-management of CFD. There was a complex interaction between balancing blood glucose, insulin and nutrition; each component was related to, and directly influenced by, each other.

#### 4.2.2.1 Insulin – understanding and uncertainty

The use of insulin posed many self-management questions, with how much insulin is needed being the most daunting. There was a sense of uncertainty, loss and bewilderment in how to gauge the correct insulin dose. It was unclear if this was

because of lack of understanding, a reflection of the complexity of CFD or a combination of both.

Mary displayed feelings of loss and bewilderment concerning how to manage her insulin. She showed a lack of understanding about how her background insulin worked and this contributed to poor adherence with it because she didn't "*see any benefit in it because it's, 'cause it's long, long lasting*." She had limited comprehension about her insulin dosing and this contributed to her not managing her blood glucose levels effectively. She also demonstrated a lack of insight into the associated risks of not controlling her blood glucose to avoid hypos.

> *I don't know [laughs]. I don't know. I just think, 'Right'. I don't know honestly, ****. I just used to make it up as I go along, yeah, and that's, that's the honest truth and I'd rather take too little than too much because of a hypo. (Mary)*

Paul and Ana learnt how to manage their CFD through more experiential learning, reflecting the fact that CFD was changeable and tricky to manage. CFD management required individuals to be more self-directed and pragmatic.

> *I think my experience of diabetes is like there's actually no way of prescribing for it in a way. Erm, you know, there's – you can't actually just say, 'Right, this is your dose and take this and it will be fine'. It's not like, you know, with your antibiotics and it's one gram twice a day, or whatever, you know, or 'Take this three times a day'. There's no, there's no way of prescribing insulin. I think erm, that is purely – like I said, you know, you could eat the same food on a different day and you could need totally different insulin requirements. So that learning was all just getting used to myself and how I erm, kind digest those carbs and stuff like that and just then learning how to add in insulin into that. (Ana)*

> *Erm, that's always an intriguing one, slightly frustrating, er, because there are times when you just don't, I've felt I just can't quite get a grip on it or a handle on it enough and you think well why, am I doing something wrong, what am I missing. Erm, but then there are other times when you think well and you realise that perhaps actually you should have had a bit more insulin with that particular food and you correct it and then the next time you eat that particular food and test your blood sugars afterwards, you've cracked it, your blood sugars don't rise as high. (Paul)*

There was recognition that insulin should be taken before food, although this was not always easy to achieve, and injections were given at different times due to forgetting,

hunger and not wanting to inject in front of others. Some undesirable consequences of taking insulin at incorrect times were observed, these included the direct impact on respiratory symptoms and blood glucose levels.

> *Sometimes I'm forgetful and, you know, I'll have my insulin at the end of a meal, that seems to be quite, erm, common recently, erm, and I do, you know, as a result I will be quite tight chested for however long and then my insulin will kick in. (Karen)*

> *if you're out and about some people will just do it in front of people I'm much less comfortable doing that I'd rather take myself off to a loo or something private and if that's not accessible then I will wait until I can get to a place where I can do it privately which isn't great because that's when erm if I've eaten then I have to wait before I do my insulin and stuff like that. (Jane)*

Ben understood that he should take his insulin before he ate so his insulin "should be kicking in as the food is being digested." A reason he attributed to not taking his insulin was the minor inconvenience of having to go and get it from "his bag," he felt silly saying it, but stated "my food's right in front of me and it's hot and I'm hungry." Another influencing factor on Ben's adherence was that Ben saw "no real immediate ill effects" of not taking his insulin, suggesting he would not "feel rubbish" if he doesn't take it. However, Ben was insightful that "*the long terms effect will be bad."* Demonstrating that having the knowledge and understanding doesn't always result in doing.

#### 4.2.2.2 Blood glucose monitoring- comprehension, confusion and compromises

Ana felt that despite being a "big thing" blood glucose testing was essential to understanding blood glucose and "yourself." Blood glucose levels were checked to inform treatment decisions regarding insulin dosing and as part of the experiential learning process to try to gain an understanding of what was happening to blood glucose levels.

> *So I take my er, blood sugar readings when I – just before I take the NovoRapid, just to gauge how much I should be taking. (Ben)*

> *doing lots of testing, I think, is quite key because, you know, like... even if I'm – you know, if I'm about to have a sugary snack, I would absolutely test beforehand just to see where I am because, you know, it's the difference – if you*

*start off with being, you know, five or if you start off being 7.5; that will be – you know, maybe a unit in the difference of how much NovoRapid I'd take erm. (Ana)*

Participants demonstrated a lack of clarity when and how often blood glucose levels should be checked. There was a lot of guilt and downbeat association surrounding blood glucose monitoring with self-judgement about their own expectations common; this included: *"I know I could do better," "not good at," "not great with," "I know I should be doing,"* and *"very guilty of."*

In the management of CFD there is more of a focus on post-prandial BG because most people with CFD lose their post-prandial response first. These were the blood glucose readings that were much more difficult and trickier for participants to perform.

*I'm not very good at, erm, taking my sugars after. I always do it before I've eaten. Two hours after, erm, I might have gone out and left it indoors. I don't necessarily take my kit everywhere with me, you know, so I'm not very good at the post blood things, do you know what I mean? (Mary)*

*it's fairly quick, it's fairly easy to do. I think the difficult ones to remember are the two hours after a meal, erm, remembering to do it pre-meal was easy, dead straightforward because you've got to sit down and think … (Paul)*

*..I know I could do better erm I do check my blood probably about four times a day but I know I would do better if I was doing it more particularly after meals, I'm good at doing it before meals but not so good at doing it after meals. Erm and that's probably, possibly why I do yoyo because I don't know what's happening after I've eaten whether it's been too much or too little or whatever*. (*Jane)*

Checking blood glucose levels was an intrusive and visible part of diabetes management; it was the one element of care which was compromised the most. Selective adherence formed a part of CFD management with choices being made regarding if and when to check blood glucose levels. With CF treatment often being prioritised over blood glucose monitoring.

*like when I do remember I will do it but it's the one thing I feel like I can, erm, be ever so slightly more relaxed on because I've got such a hectic schedule. I do need, like, I do need to, erm, you know, have a little bit of flexibility and that's where I choose to… (Karen)*

For Brian the "pressures at home or work" interfered with his blood glucose monitoring and he suggested when things get busy "doing my BMs- it's almost like the first thing that drops off the radar."

Karen's argument for not checking her blood glucose levels very often was because she felt "very instinctive" in sensing what her blood glucose were.

> *Like I'm, if I'm high I will know and then I will sometimes check, double check to confirm that and then if I'm, I, I can tell if I'm even the slightest bit low, so. (Karen)*

There was some uncertainty with regards to who blood glucose levels were checked for. Tom expressed that he only checked his blood glucose "when I've got the diabetic clinic" demonstrating he was checking more for the healthcare professional than himself. He argued that testing more felt like "*one compromise too many" in his life,* having already made the compromise to check prior to his CFD clinic.

#### 4.2.2.3 Managing nutrition – the challenges and frustrations

Optimising nutrition is one of the primary goals in the management of CF. For some participants nutrition had always been a problem and they faced difficulties in balancing CF and CFD to improve their nutritional status. Brian suggests "the biggest conflict was when it came down to the, the high calorie diet," a concept he found "just really frustrating." As a consequence of adjusting dietary intakes for CF, participants had to pay more attention to their blood glucose levels and insulin doses because if blood glucose control was not optimal increasing weight was seen to be difficult.

> *I think the biggest thing was, yeah, I was so conscious that I needed to put on weight and I couldn't do it if my sugars were just all over the place. So it was – yeah, that was just kind of something that was quite frustrating, I guess. (Ana)*

> *the last clinic my weight was down a little bit, could have put a bit more weight on again so upping the diet, increasing the calories invariably means you know more blood sugars to have to then think about as well. (Paul)*

For Jane a "lack of appetite" caused further concern to her and she required oral nutritional supplements. She described these as "really sugary" and they had a knock-on effect on her CFD and caused her blood glucose levels to "yoyo." This resulted is her

having to keep "a much closer eye" on them. There was a sense of frustration for Jane in trying to balance out the benefits of the nutritional supplements with the negative effects they have upon her CFD.

> *Yeah, yeah particularly when I'm having to have such high sugar supplements I think it was so much better before I had to rely on those but because I have to rely on them so much it's not just one or two a day its really kind of trying to pack them in that, that just throws you all over the place. (Jane)*

Jane's experiences of managing her nutrition were exceptionally demanding and frustrating. To a point she suggested, albeit in jest, "*I would have great levels if I didn't eat [laughs] and it's the eating and things that seem to throw them into chaos."*

Over time Mary began to realise, and demonstrated understanding, that her diet in the form of snacking, especially nocturnal snacking, contributed to her high blood glucose.

> *I eat during the night as well and I could get up, wake up and think, 'Ooh, I'm a bit hungry. I'd like one of them'. And I'll go and eat a bag of crisps, you know, me chip sticks or a cream egg or something and I'll go back to bed and fall asleep, and, you know, that's it. So I wake up in the morning, my levels are high so I've got to get out of that snacking.*

This enabled her to identify that she needed to make changes to her diet. Mary suggested "*I can't snack and eat as, like I used to and everything. Like my, erm, levels are always high, always."*

Tom reflected on changes he made to his diet from both a CFD and health perspective. He had *"cut down"* on crisps, fizzy drinks and sweets but described his weakness as *"stupid gummy sweets,"* which he *"used to love*" but now only has as treats for example when at the cinema. Tom was not upset and did not demonstrate a sense of loss for these foods, part of which could have been facilitated by the choice being his and he still allowed himself treats. There wasn't a sense that it was difficult for Tom to have made these adjustments.

Brian didn't let his CFD get in the way of his eating, instead he made changes to his insulin. He suggested "*I don't stop myself eating anything that is really sugary because*

*I just inject a bit more and that is, I suppose, a way of combating that and managing it."*

The role and appropriateness of carbohydrate counting in the management of CFD is not fully established and experiences were mixed. Learning about carbohydrates was individually driven and required time, self-motivation, confidence, support and a desire to gain a better understanding.

Through experiential learning Ana and Karen had become accustomed to how different foods affected their blood glucose levels, so they no longer routinely carbohydrate counted.

> *I don't carb count, per se, but I've good idea like, you know, what kind of effect certain foods will have on my blood sugars and kind of act accord-, accordingly. (Ana)*

Ben suggested that he wasn't going to embrace carbohydrate counting as he felt his CFD didn't affect him "*greatly enough to, to do maths [laughter]."* And, he "hates maths*."* He also demonstrated a reluctance to take on the additional burden of tasks and interference in mealtimes that carbohydrate counting, would mean to him.

> *it's enough faff for me to remember to take my insulin once there's a hot meal in front of me to then think about calculating carbs is something I'm not going to do when I'm about to enjoy a meal. (Ben)*

Despite Brian's acknowledgement "that I need to start looking properly at that" he described a lack of confidence in trying to start and engage with carbohydrate counting. For Brian this could be another reflection of the shame in having had CFD for a long time and not being able to ask for help.

Jane found learning to carbohydrate count helpful, it wasn't difficult for her and she learnt it *"*pretty quickly*."* It contributed to increasing her knowledge and understanding of the roles of insulin and carbohydrates as she didn't realise that there was "a link between how much insulin you have with the carbohydrates*"* prior to receiving advice*.*

### 4.2.3 The unmet need for information and support

This superordinate theme explored what information and support was needed and how it should be provided.

#### 4.2.3.1 The need to feel informed, skilled and supported

Participants were aware that CFD was not the same as type 1 or type 2 diabetes, but demonstrated little insight or understanding as to why this was. This led to feelings of uncertainty and wanting to "know why it's different."

> *And also I've been told from the very beginning CF diabetes is different from diabetes 1. How is that? I don't, you know, 'cause obviously everyone that I know that has got diabetes, they, you know, it's different and I don't see how. (Mary)*

For Ana the barrier to understanding the difference between type 1, type 2 and CFD was in being able to explain this "effectively to other people" to prevent their "judgement." She gave an example, which to her was amusing, concerning the dietary differences of CFD which was one of the biggest conflicts between CFD and type 1/ type 2 diabetes. This reflected how society portrays diabetes particularly the influence of people's ideas of what food is suitable for people with diabetes.

> *You're like, 'Oh, I'm diabetic' and then you eat cake and you're like, 'It's fine. I've got CF diabetes. It's okay' [laughter]. (Ana)*

Participants' knowledge of the impact of CF on CFD and vice versa was limited, it was difficult to ascertain if this was because they have not been told this information or if it was something they didn't understand. Karen was very insightful of the harmful link between high blood glucose levels and "bacterial infections" in the lungs however she acknowledged this was unfortunately not commonly known amongst others with CF. She suggested *"a lot of people on this ward that I've spoken to, you know, friends of mine or whomever, they, they are completely unaware of the link and it is, it is integral."*

Mary was aware of the link between CF and CFD which she represented as a vicious cycle. This could partly be because it is what the medical team told her it was, but it also illustrated what CFD was to her. This cycle to Mary symbolised CFD; because she

was not coping or managing CFD well and in her eyes not being supported. There was the representation of just going around and around this cycle, with no way out.

The impact of high blood glucose was a common occurrence amongst the participants. Tom was the only person who wanted to understand about any relationship between low blood glucose and CF.

> *actually maybe more information about – in terms of the impact of er low blood sugars on, on general health would be good, in terms of how they interact with CF and vice versa would be good. (Tom)*

People with CFD felt the need to be more supported to help alleviate the general feelings of uncertainty including the lack of knowledge and experience to *"know if you're doing it right."* There was a need to be supported by healthcare professionals experienced in CFD management. Brian was diagnosed with CFD as an adolescent and supported by a paediatric type 1 diabetes nurse. Reflecting back Brian demonstrated doubt that she was experienced in CFD management and there is a sense that he hadn't received the specialist care to meet his needs.

> *although it was a Diabetic Nurse, she didn't really – I'm sure she knew about CF but it wasn't her speciality. It wasn't her forte, so her background was diabetes and that was very different, and she was used to treating for the children that she saw. They were Type 1 diabetics that, you know, had a completely different er, complexity about their condition, erm... (Brian)*

Support to manage CFD was more than just being told what to do. It was about learning and developing a level of understanding. Karen described the understanding of why treatments needed to be done as pivotal in the management of CFD and that it was "slightly condescending" to not be informed. She suggested that increasing understanding "will empower patients to want to take better care of their health if they know why they need to do whatever they need to do."

For some individuals support was not just providing education and assistance to develop self-management skills it was also about providing a balance between educational support and emotional support to help the development of coping skills to be able to manage the additional demands of CFD. This was reflected in Mary's

struggles with CFD where she was not receiving appropriate support to meet all her needs, which left her with feelings of frustration and anger.

> *I wish that I knew more about how to look after it instead of being told how to do it. I wish I had more care about, erm, how to cope with it, that'd be one big thing, how to cope with it and that, erm, and, and more support of how you get on with it and, and, er, yeah. (Mary)*

The acquisition of skills in all three components of the CFD self-management triad (insulin, nutrition and blood glucose) were essential to the effective management of CFD. Blood glucose monitoring was considered the most intrusive form of management and the most likely to be compromised; there was the desire for practical and realistic advice. Tom, the only participant on once daily insulin, checked his blood glucose levels just prior to attending clinic appointments. He wanted to know "the minimum you should, you could get away with, from the point of view of it's useful for the doctors to know." His view on blood glucose testing were more about providing information for the medical team than guiding his management. This demonstrated his questionability in the values of checking blood glucose when only taking basal insulin and provided an insight if different perspective on blood glucose monitoring according to treatment type are required.

More support in helping participants with the challenges of trying to understand the relationship between their blood glucose and what it means in terms of their insulin doses was required. For Mary this was about being seen as a person with a life and not just a source of numbers that represented her CFD control. She desired some insight from healthcare professionals about the impact of CFD upon her life. She was angry and irritated about the way that she was treated in her consultations, this affected her self-worth as she needed more positive reinforcement to support her and to acknowledge her efforts.

> *IV* *[…] I think, like I say, in clinic you're being told to do with your levels to change them, this, that, and rather than someone helping you. There's a difference between being told what to do rather than someone sitting there helping you, yeah.*
>
> *I:* *So what's the difference?*

*IV: 'Cause they're looking at, they're looking at numbers on a piece of paper whereas you're looking at it as a way of life and doing your best [sighs].*

Information and support regarding insulin management also needed to focus upon what individuals should know to effectively self-manage their CFD. This will help increase understanding and includes: how insulin works-both "physically and biologically," what is the optimal time to take insulin and why, and how and where to administer it.

The need for dietary information was more individualised and depended upon nutritional status. How to manage meals was difficult for some. Jane was uncertain how to manage her meals times demonstrating a lack of knowledge and ability.

*Erm mmm how to manage meal times cos I still don't think I have that down because I still when I eat seem to go up really high. I don't know if that's normal and then I'll come down straight away. (Jane)*

There was uncertainty if information on carbohydrate counting would be beneficial or not in the management of CFD. Brian felt that support on carbohydrate counting "would be a really good tool to help someone with diabetes manage their condition better," however he has generalised this to people with CFD and not personalised to himself. This could further reflect his lack of personal engagement seen earlier.

#### 4.2.3.2 Creating systems to deliver trustworthy information and support

Peers and healthcare professionals had an active role in providing information and support to people with CFD. The use of web based social media for communications such as Twitter or CF forums was generally helpful in these circumstances.

*I think it's a really good mechanism to get people that can't meet face-to-face to actually talk and share problems and share solutions. (Brian)*

Participants found "reassurance" in knowing that other people with CF were also experiencing similar signs, symptoms or experiences to them. Ana felt that this helped to "normalise" experiences.

*[…] nice to know that other people have experienced something that you're experiencing and, and... to get their take on it and, and to know what helped them. (Ben)*

*So I think having that kind of ability to interact with other people – I think it just probably normalises your situation and then that kind of just reassures you but yeah, there were other people who had had similar erm, (Ana)*

*By giving you different ideas about how to cope, letting someone know that, you know, how they think and that they're going through the same and, you know, it's just something like that. (Mary)*

However, there was a need for reliable and respected peer support from trustworthy websites and forums and by establishing relationships with "the people whose opinions you trust."

*So you get to know the people whose opinions you trust; erm, who you know are not hyperbolic and erm, who, you know, you can actually kind of – you share those ex-, - kind of same views and values with. (Ana)*

On the negative side, due to the variability in the clinical course of CF, not everyone using the forums or social media were at the same stage of disease. For some this was depressing and led to unhelpful thoughts about the future or brought up emotions that people were not ready to explore.

*I did look at forums once or twice and just found a lot of very morbid, negative sort of comments […] (Paul)*

*I think because there's a lot of people, you get some people who are really into it but they might be further along in their condition than you and they talk about things that are potentially in your future and you just don't wanna, don't wanna be there that yet. Its erm its too heavy… (Jane)*

Ways to provide information included the use of written information, videos and via the Internet. The usefulness of leaflets was limited; they tended not be highly valued and contained more generic rather than specific advice. There was the questionability of "do we read them?" and a sense of leaflet fatigue as they were provided by most healthcare professionals. As highlighted by Paul the benefits of leaflets were short lived.

*I think they're initially helpful, erm, because you've got something there tangible to actually to deal with. Erm, however they, then you end up sticking them in a folder or they get lost in a pile of paperwork somewhere so then there needs to be something longer term to go back to as a reference point. (Paul)*

Information on the Internet was deemed to be more preferential than written information in leaflets. It was felt to be more helpful by alleviating the issue of storage and by being more up to date, relevant and long term.

*Erm, but obviously you want something that's going to be easily accessible longer term so be it on a website or something that's just, as a reference point for when the leaflet gets lost or forgotten or something like that. (Paul)*

*So I think it's always good to have something that you can refer back to but written literature on a website is probably going to serve a better purpose than a leaflet that you'll take home, forget where you've put it, find it in two months, read it and go, 'Oh yeah. (Brian)*

All participants, except Mary, were happy to "Google" information on the Internet for themselves. Mary did not view this as beneficial and she felt it was not something she should be doing. For Mary there was a mismatch between what information was needed for education and what was needed in terms of 'what is help for her,' which may have been emotional support.

*You know, it's not something that I think I should get off me arse and look at meself, I think someone should be helping me how to cope with what I'm going through. (Mary)*

Videos as a source of information were generally felt to be helpful, particularly for people who learnt in a more "visual" way. However, to be beneficial they needed to be "relatively short" to "ingest." Other CF team staff had already produced videos for patients and the experience of using these was "really useful" and helped to go back and "revise" what had been taught. For people starting out with CFD videos concerning giving insulin and testing blood glucose were suggested.

*here's a few videos on YouTube to watch about, you know, the pen, the insulin and putting a cartridge in, and this is a BM machine, and this is how you do it, and this is...' you know, little things that you could visually see you know, watch the video erm, see it happen and sort of get to grips with it yourself. (Brian)*

*Erm, so I don't know whether it's, yeah, kind of videos around, I guess, for people starting out; like how to test and how to inject yourself erm, and what you need to do and stuff like that. That would be probably useful. (Ana)*

Healthcare professionals were the primary point of contact for people with CFD and a trusted source of advice and hence held a very important role in the management of CFD. The recognition of "professional first-hand" advice, which was aimed at the individual was highly regarded, with more impact than just "reading" something for yourself. However, not all participants were getting the correct sort or level of support they needed.

Ana needed support and "reassurance" to assist her adjusting her insulin dose, so she felt "safe."

*[ …] you know, talking to somebody at a clinic or, you know, even a phone call erm, just to be like, 'Yes, you're doing the right thing'. (Ana)*

Karen was angry because she didn't feel supported and there were discrepancies between what she perceived support to be and what she received.

*Not, I don't think what they class as support is necessarily, I don't think it's support, I think it's just bombarding you with, you know, printouts of regurgitated, outdated information. (Karen)*

Mary was frustrated and annoyed that she was not getting the type of support she needed; this was a source of distress for her.

*Well with a bit more help and motivation. You know, it's all right sitting there, someone telling you what to do because then I get angry and think, 'Well, you know, fuck you. You're not going through it', do you know what I mean? (Mary)*

The CFD clinic was a limiting factor in provision of support due to time commitment and barriers caused by the provision of separate CF and CFD clinics. Time was in terms of how much patient's time it required to attend an additional clinic. Jane was angry and resented attending CFD clinic because the time of her journey significantly outweighed the benefit she saw of attending the clinic because "nothing changes."

*I do find the appointments quite difficult [..] it will take me an hour and a quarter, hour and a half to get here then I'll be in there for 20 minutes and then*

*I've got the whole journey home. So, which erm so it often feels like I've come in for nothing because a lot of the time nothing changes erm so I'll come in they'll ask me a few questions and then and then that's kind of it. So, it does feel like it's a lot of effort to have to, to do the diabetes appointments as well that is yeah. (Jane)*

Time was also a reflection of how infrequently CFD clinics were held. Mary was irritated about this; she felt the service provided was not supportive of her needs.

I *think, erm, just a bit more frequent than every bleeding six months when you can get an appointment in a diabetes clinic 'cause they're not helping you they're just telling you what to do. There's no help there, is there? (Mary)*

Suggestions to improve CFD support included treating CFD and CF "harmoniously" as they were currently being "treated separately." Incorporating CFD care into CF clinics was suggested to: improve efficiency, increase attention paid to CFD management and integrate CFD management into routine CF care.

*I think it'd be good to get, yeah, and I think that's where having someone in a CF clinic – a diabetes specialist in a CF clinic, to talk to CF related diabetics would be good. And you'd be able to get more out of that I think. (Tom)*

*Erm that would be nice if the two could be combined just to, I don't know, I know erm logistically that probably would, is difficult but it just reduces given the amount of erm trips these days anyway I'm having to make to the hospital it erm would just be erm yeah. (Jane)*

## 4.3 Discussion

The key findings, from each superordinate theme, are discussed in relation to the research question "what are people's experiences of self-managing their cystic fibrosis diabetes?" findings from the meta-ethnography (chapter three), current literature and theories. Clinical implications of the research findings, methodological issues, ideas for further research and study reflection will then follow.

### 4.3.1 Forming a relationship with CFD

This superordinate theme was divided into three subordinate themes: CFD- impact, constancy and exposure, the conflicts, complexities and unpredictability of CF and CFD and facing morbidity. Essentially the core of the participants' experience of forming a

relationship with CFD appears to be around developing a balanced relationship between CF and CFD.

#### 4.3.1.1 CFD-impact, constancy and exposure

The development of CFD had both physical and emotional impact on the lives of people with CF. Attitude and perception of CFD helped facilitate CFD management but the constancy of it posed one of the biggest challenges. Developing CFD heightened the sense of exposure through illness visibility.

Participants described feeling a range of emotions, mostly negative, when diagnosed with CFD. This led to uncertainty and confusion especially with regards to the significance of having CFD. This is consistent with experiences of people with type 1 diabetes who also reported negative emotions such as confusion, sadness, shock and fear upon diagnosis (Schur et al. 1999, Celik et al. 2015, Robinson 2015, King et al. 2017) and seen in people with CFD (chapter three). Age at diagnosis was an important consideration in the adjustment to having CFD. Adolescence is a particularly challenging time due to physical and psychological changes and the greater emphasis placed on self-caring. Half of the participants, in this study, were diagnosed with CFD during adolescence; they all acknowledged this was a challenging time emotionally and physically, but the majority took CFD within their stride.

Maintaining a positive attitude and viewing CFD as part of CF facilitated acceptance and management, and limited the impact of CFD. Where CFD was viewed as part of CF then management tasks were incorporated into established CF routines. All but two participants accepted their CFD and reported just getting on with it. People who develop diabetes have to learn how to live a new way of life with diabetes (King et al. 2017). Time is an important factor and gradually over time people with diabetes become able to incorporate having diabetes into their sense of self (Robinson 2015). Limiting the impact of and adapting to CFD involved the process of normalisation, this is recognised as a basic strategy used in people with chronic illness to deal with the challenge of their illness (Kelleher 1988). Seeing oneself as ‘normal’ or getting on with life is a highly regarded aspect in the lives of young people living with chronic illness

(Taylor et al. 2008). For people with CF this is about trying to achieve a sense of balance between self-care activities and daily life.

Control over diabetes is a fundamental aspect of management and is an important feature of adaptation and coping (Schur et al. 1999). People with diabetes aim to be in control; in order to cope, they attempt to control diabetes so that it fits in with, rather than dominates, their daily lives (Freeborn et al. 2017, King et al. 2017, Nishio & Chujo 2017). Being unable to cope with CFD was linked to not accepting it and trying to manage it as an addition to CF, thus increasing treatment burden and management activities.

On its own CF is a disease that requires attention to time, planning and organisation but it was the constancy and invasiveness of CFD into daily life that was considered a barrier to its management. Participants needed to plan ahead and be organised- CFD was always there and there was therefore a loss of spontaneity. The inconvenience and inference of diabetes, particularly with regards to administering insulin and monitoring blood glucose levels, is also a challenge for people with type 1 diabetes (Freeborn et al. 2013, Celik et al. 2015, Robinson 2015, Jull et al. 2016) and thus not unique to the management of CFD.

Developing CFD increased the visibility of CF. Many people with CF chose not to disclose about their illness, they carry out most of their treatments in their own homes and disclose on a need to know basis. People with CF choose carefully who to tell about their CF to avoid being judged or stigmatised (Gjengedal et al. 2003, Borschuk et al. 2016). They modify their behaviours in order to minimise the impact of any treatments, such as pancreatic enzyme replacement therapy, which are required on a more frequent basis and thus need to be taken outside the home. CFD requires treatments and monitoring that are more difficult to hide and thus make it visible and attract attention from others. People with type 1 diabetes (Freeborn et al. 2013, Chilton & Pires-Yfantouda 2015, Robinson 2015) experience a strong sense of feeling different from peers because of their illness. Diabetes is a visible disease in terms of what is required to manage it on a daily basis. This visibility may lead to stigma- the

fear of being discovered as different and of being judged by others as less acceptable than the norm (Robinson 2015). Adaptations in behaviours occur to maintain identity as a well person, Schur (1999) terms this 'stigma management'. In this study changing habits were seen particularly around eating with others, where participants would administer insulin away from the tables to avoid drawing attention to themselves. It has been suggested that individuals have both public and private accounts of illness and, within social contexts people often try to convey an identity which is acceptable to others (Kelly & Field 1996). Thus, administering insulin or checking blood glucose levels in public to some individuals would not be considered socially acceptable to them and expose too much of the personal private self.

#### 4.3.1.2 The conflicts, complexities and unpredictability of CF and CFD

Conflicts and complexities exist in how CFD and CF care is managed and the relationship between them is portrayed. Both conditions are unpredictable in nature and this further complicates management.

Both CF and CFD are complex conditions that require life-long management and often complicated treatment regimens. The relationship between CF and CFD is hazy but a very important consideration in terms of management. Some participants saw links between CF and CFD only in terms of medical and dietary management but not in terms of CF causing diabetes or the impact of CF on CFD and vice versa. Whereas other participants recognised a relationship between high blood glucose levels and respiratory infections and the difficulty there was in trying to achieve a balance between them. These links are fundamental to management and more consideration should be given for treating CFD as part of CF and not a separate entity. When considered as one condition management was incorporated into established CF treatment regimens. These links between the two conditions echo those seen in chapter three which prompted the consideration to the name CFD.

This CF centre contributed to the further separation of the two conditions and created a barrier by having a separate CFD clinic. This service provided very little overlap between respiratory and diabetes management because diabetes issues were managed in the CFD clinic and respiratory issues in the CF clinics. This further

complicated management for patients. CF care pathways may therefore have a role in supporting people to develop a stronger notion of inter-dependence that may have life extending impacts.

The role of corticosteroids in the development of CFD and the complexities it added to the management of CFD was a significant finding within this study; it has not previously been reported as such a major concern to people with CFD (chapter three). One participant had an extremely negative relationship with corticosteroids and blamed them for his diagnosis of CFD; this led to his inability to fully accept CFD. When corticosteroids were used to treat respiratory symptoms difficulties in managing blood glucose levels were described; there was a sense of loss of control and more difficulties and uncertainties in trying to gauge insulin doses. Participants did not feel that they had been prepared enough by healthcare professionals for the consequences of corticosteroid treatment or supported enough throughout the treatment.

Living with CF is unpredictable due to day-to-day variability in clinical symptoms this can affect blood glucose levels and ultimately the management of CFD. Living with this unpredictability is challenging. People with CFD had to continually make treatment and management decisions that changed frequently. CF is a progressive illness: as health begins to deteriorate then life can start to become more restricted and treatments more complex and demanding, increasing treatment burden further. This can impact on quality of life as the severity of disease increases (Gee et al. 2000). Managing CFD was also tricky and challenging and for some it has a sense of unpredictability which further added to the complexity. Reasons for this are not clear as numerous factors affect insulin secretion and sensitivity.

#### 4.3.1.3 Facing mortality

There was awareness amongst participants that complications of CFD exist, with primary concern being for eyesight. Thoughts for the future were however predominated by CF, including the understanding that they would succumb to CF first. This led to prioritisation of CF management and sometimes compromised the treatment and management of CFD. In individuals with co-morbidities life needs often took precedence over diabetes (Wilkinson et al. 2014). Fears and concerns over the

long-term effects of diabetes has been seen in people with type 1 diabetes (Dickinson & O'Reilly 2004, Celik et al. 2015) however diabetes was the only chronic illness these participants had. CF was the most serious condition to participants in this study; there was recognition that it was a progressive life-limiting disease and therefore the long-term effects of this took precedence over CFD. However, with increased life expectancies people are likely to be living with CFD for longer. Thus, the importance of glycaemic control is exceedingly important to prevent a rise in diabetic complications and associated demands on health services.

### 4.3.2 Balancing the CFD self-management triad

CFD management aims to achieve a balance between keeping blood glucose as near normal as possible/controlling symptoms with preventing respiratory and nutritional decline. The self-management of CFD focussed around three interlinked components: insulin, blood glucose and nutrition.

#### 4.3.2.1 Insulin- understanding and uncertainty

Uncertainty regarding insulin use was problematic. This concerned how much insulin was needed and the most appropriate time to take it. This reflected insufficient knowledge and therefore demonstrated inadequate understanding of insulin and how it worked. There was acknowledgement that insulin dosing was not fixed and needed to be adjusted according to dietary intake, changes in treatments and clinical status. However, this was a dynamic process which required knowledge, motivation and support to sustain. The unpredictability of CFD itself made insulin dosing challenging and difficult to manage.

The use of carbohydrate counting to inform treatment decisions forms an integral part of type 1 diabetes self-management programmes and has been shown to be an effective method of management (Edwards 2015). However, the use of carbohydrate counting amongst people with CFD within the study was variable. This could be for many reasons. First, there are no formal structured self-management education programmes for CFD. Secondly, people with CF are taught about dietary fats from a very early age to help manage pancreatic exocrine insufficiency and the need to also know about carbohydrates is too much for some people. Finally, carbohydrate

counting requires adequate numerical skills, which not everyone possesses (Marden et al. 2012). Due to the integral part of carbohydrates in diabetes management there is a need to for people with CFD to receive some education to increase carbohydrate awareness, to help inform decisions regarding insulin dosing, and for some this may be carbohydrate counting. Thus, achieving a balance of what is the most applicable to each individual with CFD is a more appropriate approach.

#### 4.3.2.2 Blood glucose monitoring- comprehension, confusion and compromises

Blood glucose monitoring is an essential part of the management of CFD; it is a key part of the experiential learning process to develop knowledge and understanding. It was used to inform self-management decisions such as insulin dosing and to monitor for changes in blood glucose levels due to illness or lifestyle factors. However, there was a lack of clarity regarding when and how frequently to check blood glucose. It was the part of self-management that was compromised the most.

Blood glucose monitoring posed many challenges for people with CFD, in this study, with the invasiveness and inconvenience being the most predominant. This is not limited to the management of CFD as it has also been reported in people with type 1 diabetes (Freeborn et al. 2013, Robinson 2015, Jull et al. 2016). The need for post-prandial blood glucose levels in the management of CFD caused additional challenges as these were the blood glucose which were found the most difficult to check, predominately due to untimeliness. Participants in this study made informed choices regarding blood glucose monitoring which included making compromises and for some prioritising CF treatments. Not checking blood glucose levels is also common in the management of type 1 diabetes (Celik et al. 2015, Chilton & Pires-Yfantouda 2015).

There was conflict and confusion between what healthcare professionals recommended with regards to the frequency and timing of blood glucose monitoring and what participants understood and were actually doing. Tom, who was on just basal insulin, only checked his blood glucose levels prior to his CFD clinic appointment; which was annually. He did not use his blood glucose readings to inform his treatment decisions or to review trends in his blood glucose levels but just for the healthcare

professionals to review in clinic. It could be argued that his need for blood glucose monitoring is less that someone on a basal bolus regimen and he would not use it to inform decisions around meals as this is not the mode of action of basal insulin. However more individualised recommendations appropriate to his needs were required. Conflicts between blood glucose monitoring recommended guidelines, healthcare professionals' expectations and patients' motivation have been seen in people with type 1 diabetes (Celik et al. 2015). Where there is a trend to check blood glucose 'passively' to please others rather than as a self-management tool (Celik et al. 2015, Chen & Chang 2015). Thus, self-management of blood glucose levels needs to be seen as being of benefit to people with all types of diabetes. A sense of balancing blood glucose monitoring into their lives needs to be achieved. For people with CFD other routine treatments and management needs to be considered and balanced with when and how frequently blood glucose levels need to be monitored.

#### 4.3.2.3 Managing nutrition – the challenges and frustrations

Achieving and maintaining optimal nutritional status is a key part of the management of people with CF; the addition of CFD presented further challenges to this. Seeing a dietitian forms part of routine CF care; people with CF are assessed and given dietary recommendations appropriate to their individual needs (Cystic Fibrosis Trust 2016).

Managing nutrition was a challenge to the participants within this study. The need to further increase dietary intake through food or oral nutritional supplements further exacerbated this. Making these dietary changes led to further attention needing to be paid to blood glucose levels and adjustments in insulin doses to achieve balance. Difficulties in dealing with dietary aspects of CFD and balancing it with maintaining weight has been previously reported (Lake 2010). Managing nutrition is not just a problem in people with CFD; in one study of young adults with type 1 diabetes diet was deemed the biggest problem. These young adults had knowledge of what not to eat and when to eat but some of them did not have knowledge of carbohydrate counting (Celik et al. 2015).

To manage CFD some participants chose to make modifications to their diet. These included reducing carbohydrate intake, missing meals and limiting snacking. With

improvements in nutritional status seen in people with CF is it no longer unusual for some dietary modifications to be recommended for managing CFD, however in this study these changes were decided by the individual. It is understandable that some individuals may want to snack less to avoid additional insulin injections and to eat less carbohydrate to reduce the amount of insulin required. This highlights the importance of healthcare professionals assessing the appropriateness of this on an individualised basis. In this study there wasn't a sense of dietary changes being detrimental to health, they were individual's choices in how they wanted to manage their own situation.

As previously discussed, the use of carbohydrate counting in the participants' management of CFD was variable. The appropriateness and benefit of carbohydrate counting for people with CFD is not fully established although some participants in this study have found it helpful. Currently carbohydrate counting tends to be taught on an ad-hoc basis and not to form part of routine education (Cystic Fibrosis Trust 2016). This may be because there are no established self-management education programmes for the management of CFD.

### 4.3.3 The unmet need for information and support

#### 4.3.3.1 The need to feel informed, skilled and supported

As also demonstrated in chapter three there was a lack of knowledge and understanding concerning the differences between CFD and type1/ type 2 diabetes; participants were aware that it was different but were uncertain how and why. There was also a lack of understanding of the impact of CFD on CF and the physiology of CFD. This is not unique to CFD as people with type 1 diabetes have also expressed a desire to know more about the physiology of diabetes (Roper et al. 2009). Thus, there is a need to provide people with CFD information about what CFD is, what causes it and how it impacts on their CF and hence their health and well-being.

Participants demonstrated limited knowledge of the relationship between the impact of CF on CFD and vice versa. Mary's representation of this relationship was as a vicious cycle. However, Mary was struggling with management and the cycle representation only further added to her anxiety as it was difficult for her to see a way to get out of this cycle as she was not coping and not, by her definition, being supported. People

should be informed about this relationship from diagnosis, to help inform management decisions.

Education and support regarding the CFD self-management triad needs to be practical, realistic and helpful. This should include: recognition of what is achievable in terms of blood glucose management, advice on managing insulin, clearer information how to achieve good CFD control whilst also maintaining weight and increased carbohydrate awareness. What is important is that individual needs are assessed and accounted for when tailoring education and advice. This was also identified as essential for people with type 1 diabetes (Wilkinson et al. 2014, Celik et al. 2015, Jull et al. 2016).

A general lack of support to participants with CFD in this study was observed. Clearly this gap in service provision needs to be addressed and more support provided to facilitate the self-management of CFD. However, attention needs to be paid to the type of support provided. With the need for educational support to help develop knowledge and self-management skills and emotional support to address the psychological impact of CFD. Participants want support from people who have knowledge, understanding and experience of CFD. Collaborative planning of care between healthcare professionals and people with type 1 diabetes has led to feelings of support (Spencer et al. 2010) therefore, actively involving individuals with CFD into their care planning may be one way to facilitate support and achieve the sense of feeling supported.

#### 4.3.3.2 Creating systems to deliver trustworthy information and support

The segregation of people with CF due to infection control policies within CF centres prevents face-to-face peer support (NICE 2017). This has been shown to create a sense of isolation in people with CFD (Tierney et al. 2008, Richmond 2012). Despite segregation not being an issue, feelings about being alone have also been seen in people with type 1 diabetes (Celik et al. 2015, Robinson 2015).

The participants with CFD in this study did not report a sense of isolation or being alone except for Mary who gave the impression of struggling alone, with insufficient support. Some participants turned to social media and the Internet for support with

their CF. This however needed to be from a trusted source to be considered helpful. Lack of control of information online is a disadvantage of this means of support (Kirk & Milnes 2016). Pearson & White (2014) conducted a study examining nutritional queries, over a 6-month period, on the online forum of a CF charity. They found that CFD was the most frequently raised topic; with a lack of understanding of it regularly identified (Pearson & White 2014). Online support groups have been shown to enable young people living with CF to share their experiences and feelings with their peers, this can support their self-management and help empower them in their interactions with healthcare professionals (Kirk & Milnes 2016).

The use of the Internet was considered a more preferential medium for information than leaflets. With all but one participant happy to source information for themselves on the Internet. The use of videos was considered useful for visual learners, however careful consideration to their duration should be given. There was a preference for short videos. Some participants had already experienced using videos produced by members of the CF team which they found to be helpful.

Healthcare professionals were the first point of contact for the participants. There was, however, a need for healthcare professionals to show a greater insight and understanding of the experiences of living with CFD. There was a need for support rather than being told what to do. The management of diabetes needs to be considered in a wider context beyond the disease itself by adopting a more person-centred approach which considers social, emotional and lifestyle issues (Wilkinson et al. 2014, Celik et al. 2015, Jull et al. 2016).

The CFD clinic was a major drawback in the provision of CFD care within this centre. This was due to time, specifically the amount of time it took for patients to travel to attend CFD clinic, the infrequency of CFD clinic, and lack of synchronicity with routine CF care. People with CF already attend hospital regularly for reviews, with average outpatient appointments taking 4 hours 46minutes (Cystic Fibrosis Trust 2018b), and the frequency of contact increasing with the severity of disease; there was generally a lack of desire to attend the CF centre more than necessary. Leave from work/college

and expense also need to be considered. Alternative methods to delivering CFD specialist care which will have less impact on time and patient expenses such as remote clinics or incorporation into routine CF clinics is therefore more appropriate to meet the needs of this population.

### 4.3.4 Strengths and Limitations

This IPA study is methodologically congruent in including this small sample of adults living with CFD (Smith et al. 2009). IPA is an inductive approach and gives participants freedom to express their experiences in their own words, with a trend in this study for more of a focus on the problems of self-management rather than solutions to it. The use of IPA allowed the aims of this study to be met. A more structured methodology may have given more positive experiential ideas for developing the self-management education programme, which IPA has not fully facilitated but it may not have produced such a detailed representation of individual experiences. The stakeholder development group in stage 2a will contribute to positive experiential ideas to inform the development of the CFD self-management education programme.

This study aimed to describe experiences of both adolescents and adults with CFD. Twenty-two of the 44 invitation to participate letters were sent to people less than 26 years old. Unfortunately, only one of these responded, highlighting the difficulty recruiting this younger population. All the participants in this study had CFD for six years or more. The self-management experiences of younger adults or those with new or relatively newly diagnosed CFD may therefore not be fully reflected in these findings. The experiences of the participants, within this study, are from one CF centre and may not be fully transferable to other CF centres where variations in the management of CFD may exist. However, they are reflective of the results from chapter three which represented findings of people with CFD from six different CF centres situated in the UK, USA and Canada.

It is also important to consider that those who volunteered to participate in this study may be more highly motivated and manage their illnesses better and are therefore happier to share their experiences. Although in the case of Mary, anger and

dissatisfaction could be viewed as her motivator to participate. Thus, the findings may not be completely reflective of the whole CFD population at this centre. However, their self-management experiences and effectiveness at managing CFD were still very varied. There are limited studies looking at the experiences of CFD indicating a need for further qualitative studies exploring these experiences to be undertaken.

My role as a researcher also needs to be considered. I was previously known to the participants as a dietitian, within the CF team, who had a role in patient care. This may have led to some participants withholding information although confidentiality and anonymity was discussed and assured. However, I felt the participants spoke freely and willingly during the interviews and I have no reservations about how the interview process went.

As discussed in chapter two, IPA adopts an ontological approach of critical realism which is located within the interpretivist framework. A reality exists independent of the observer, but it can only ever be partially known and is only available through the insights and interpretations of the individuals experiencing it. There is the acknowledgment that interpreted knowledge is not totally objective, it reflects some truth about the phenomenon, but it is influenced by our own interpretations. Therefore, as an IPA researcher I become heavily immersed in the data and ultimately my interpretations had an influence upon the results. Data analysis was reviewed by all members of the supervisory team with discussions of interview transcripts, key findings and interpretations. The results were also presented to the PhD Project Management Group, consisting of lay and healthcare professionals, for their support and advice.

### 4.3.5 Implications for practice

Many knowledge deficits were identified within this study; a lack of understanding of the link between CF and CFD was a key finding. This included what caused CFD and how its management impacted CF and vice versa. Other identified knowledge deficits concerned the CFD self-management triad- the three key interlinking components to self-managing CFD. These key findings, concerning where deficits in knowledge were

identified, will be used to inform the development of a self-management education programme for people with CFD. The MAGIC programme stakeholder development group will give consideration to these findings and decide how they will be used to influence the design of this intervention. These key findings will therefore help inform the MAGIC programme's content and structure and help address the educational needs of people with CFD. Improving knowledge will facilitate understanding, empower decision making and boost confidence to self-manage. However, the educational needs of healthcare professionals were also deficient and this also requires attention. The aim of this study was to develop a self-management education programme for people with CFD, it didn't take into consideration the requirements of healthcare professionals. The MAGIC programme could be used, in CF centres, by healthcare professionals to enhance their knowledge about the management of CFD. For healthcare professional specific CFD programmes more research is required to identify their particular needs. In the future the MAGIC programme could be adapted into a healthcare version to meet these needs.

Healthcare professionals were regarded as the first point of contact for people with CFD but a lack of support or the wrong type of support was reported. This needs to be addressed, by CFD teams, so that people with CFD can make more effective self-management decisions in a helpful and encouraging environment. However, it is not enough just to tell people what to do, individuals need an integral part in planning of treatment and management and the wider context beyond the illness itself needs to be considered.

Both CF and CFD are complex life-limiting diseases and require daily treatment and management that impacts on all aspects of life. CFD should be regarded as part of CF; it is a complication that occurs with ageing and due to CF pathophysiology. The CF centre further contributed to separating the two conditions by the way its specialist clinics and services were structured. Both conditions should be treated as one; individuals should be encouraged to incorporate CFD management into their routine CF care. Quality improvement aspects of gaps in service for people with CF and CFD need addressing to improve the delivery of support, education and care. People with

CF do not want to attend the hospital more frequently than needed; time is a very important aspect within their lives. People with CF are willing to embrace new technologies provided they are from reliable and trusted sources. Consideration should be given to integrating the CF and the CFD service, with appropriate multi-specialist input, into the clinical team. Thus, care delivery pathways, including digital, need to be developed to provide continuity of care, education and support with known healthcare professionals. This will help support the integration of people with CFD in treatment and care planning.

## 4.4 Conclusion

This study demonstrated impact and importance because despite being a very common complication of CF very limited qualitative work looking at experience of self-managing CFD has been conducted. This study has increased knowledge of the treatment and self-management challenges that people with CFD experience and identified implications for practice.

The management of CFD is a complex multi-dimensional process. The findings suggest that the self-management of CFD was met with many challenges which included: trying to balance CF and CFD, receiving insufficient support from healthcare professionals and having deficits in knowledge to make appropriate self-management decisions. A self-management education programme for people with CFD is needed to address these identified deficits to facilitate self-management. The provision of appropriate and timely person-centred CFD care and support also needs to be addressed.

## 4.5 Research Outputs

Collins, S., Jones, A., Woodward, S. & Sturt, J. (2019) P302 “If I could be off them, I would” - adults with Cystic Fibrosis Diabetes experiences of corticosteroid therapy. *Journal of Cystic Fibrosis***18**, S142. Poster presentation at European CF Conference, 5-8th June 2019.

The Cystic Fibrosis-Related (CFD) self-management triad; it is about more than just numbers. Poster presentation at Royal Brompton & Harefield NHS Trust's Nursing and Allied Health Professional Research Day, 8th November 2018.

Collins, S., Jones, A., Woodward, S. & Sturt, J. (2018b) P161 The Cystic Fibrosis Diabetes (CFD) self-management triad; it is about more than just numbers. *Journal of Cystic Fibrosis***17**, S104. Poster presentation at European CF Conference, 6-9th June 2018.

PhD project management group meeting 2, 4th January 2018- presentation of IPA study

# 5 MAGIC programme development and review

This chapter focuses on the process of integrating the findings, from the previous stages of research, with relevant theories to inform the development of the MAGIC programme. This stage of the study will answer the question "what does a web-based self-management education programme for CFD need to contain?" In this chapter the methods and results for the MAGIC programme development and the MAGIC programme review will be discussed separately. The findings will then be consolidated into a joint discussion and conclusion.

## 5.1 Method - Stage 2a MAGIC programme development

### 5.1.1 Research design

This study aimed to develop the MAGIC programme as an e-learning resource. The development phase outlined by the MRC Framework supported the design of the MAGIC programme (chapter two). Hence, the development of the MAGIC programme was informed by findings from stage 1a and 1b, co-design using a stakeholder development group and relevant theories/ criteria (figure 16).

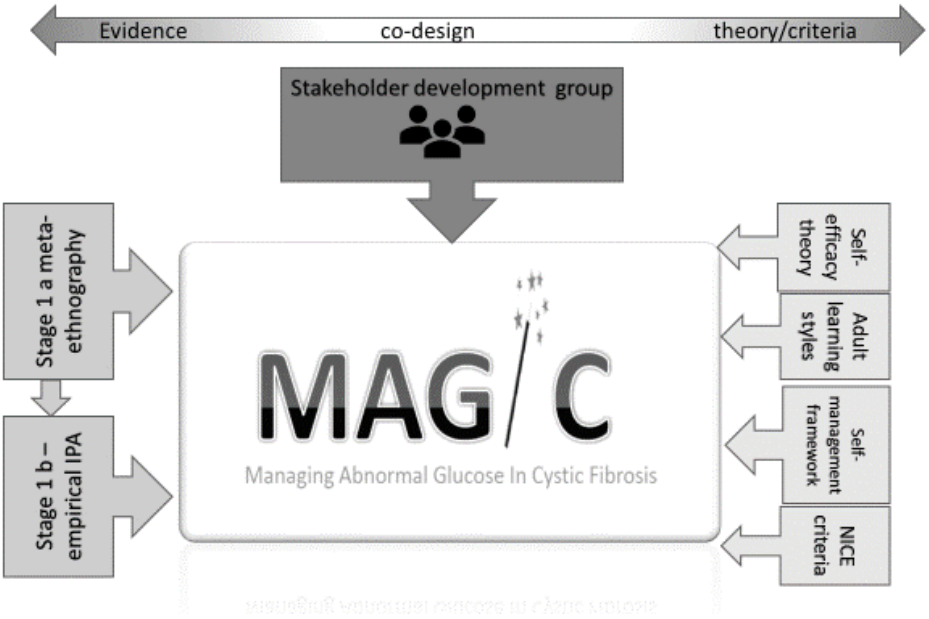


**Figure 16 Development of the MAGIC programme**

### 5.1.2 The MAGIC programme stakeholder development group

The stakeholder development group consisted of expert healthcare professionals, from CF centres throughout the UK, and people with CFD (table 24). Nine healthcare professionals were recruited through professional groups and by invitation based on their recognition as experts in CFD management. Seven people with CFD were members of the stakeholder development group; they were recruited via the CF Trust, social media (Twitter) and the CF centre; five of these attended the stakeholder development group meetings and two contributed to the group via email (e.g. reviewing modules, videos and content). These were not the same people with CFD who participated in the IPA study described in chapter four. Four other people (one parent, two people with CFD from other CF centres and a person with CF) initially agreed to be a part of the stakeholder development group but they did not respond to further contact and thus did not contribute to the MAGIC programme development.

**Table 24 Stakeholder Development Group**

| Stakeholder development group members |
|---|
| • **Project lead**<br>• **7 people with CFD (4 female)**<br>• **1 CFD nurse specialist**<br>• **1 CF nurse specialist**<br>• **4 CF Specialist Dietitians**<br>• **2 Consultant Endocrinologists**<br>• **1 CF Specialist psychologist**<br>• **Digital content manager**<br>• **Supervisory team** |

Other people who contributed to the stakeholder development group meeting included:

- Digital content manager- responsible for designing the web-based programme
- Supervisory team –expertise in DSME, chronic disease management, delivering education, cystic fibrosis and behavioural medicine. They contributed to meeting discussions, provided support and helped with taking notes during the meetings

- Student nurse on research elective- supported literature reviews on theoretical underpinnings of DSME programmes, helped with taking notes during the meetings

The stakeholder development group was regarded as a study group; by volunteering people were consenting to contribute to the development of the MAGIC programme. Ethical approval was therefore not required as these people were not regarded as study participants. No financial rewards were received for being part of the stakeholder development group, however travel expenses were reimbursed. All data presented to the stakeholder development group from stage 1b was anonymised; no identifiable data were shared.

To prevent cross infection in people with CFD we needed to use videoconferencing software to facilitate contact between group members. The efficacy, ease of use and cost of different options for videoconferencing were considered. There are numerous videoconferencing services available e.g. Zoom meeting, Adobe connect, BlueJeans, Skype, Google chat etc. I had experience of successfully using BlueJeans videoconferencing service via work as steering group member with the James Lind Alliance top ten research priorities for CF group. I was also familiar with using Skype. These were therefore the two chosen preferences considered as a starting point. On three test runs BlueJeans was found to be more reliable and easier to use than Skype; all test participants could log onto BlueJeans and participate in the video call, whereas this was not achievable with Skype. The cost of BlueJeans was reasonable and was met through the programme development budget. The decision was therefore made to use BlueJeans videoconferencing service for these meetings.

### 5.1.3 The process of developing the MAGIC programme

A series of four stakeholder development group meetings were held between January and November 2018 (figure 17). The process of co-design recognises equality in the contribution from all involved (Bradwell & Marr 2008) therefore, all stakeholder development group members had an active role in decision making rather than just being seen as information providers.

As the project lead it was my role to facilitate the stakeholder development group meetings, to create an environment that put participants at ease and encouraged them to contribute freely to discussions. I also had to ensure that perspectives were balanced and that all group members had the opportunity to express their opinions. The direction of the meetings needed to be maintained to ensure conversations were relevant and focussed on the desired outcome of developing the MAGIC programme. As the people with CFD were attending remotely it was important to ensure interactions with them were maintained throughout the meetings and that they were included in all conversations. This was done by actively asking opinions, using the chat feature on BlueJeans, which allowed participants to type messages, and identifying a member of the stakeholder development group to be a link person with the people at home. The link person monitored for questions and identified any technical issues that occurred with the videoconferencing software. Notes were taken during the stakeholder development group meetings and audio recording were made. This helped inform development and identified action points.

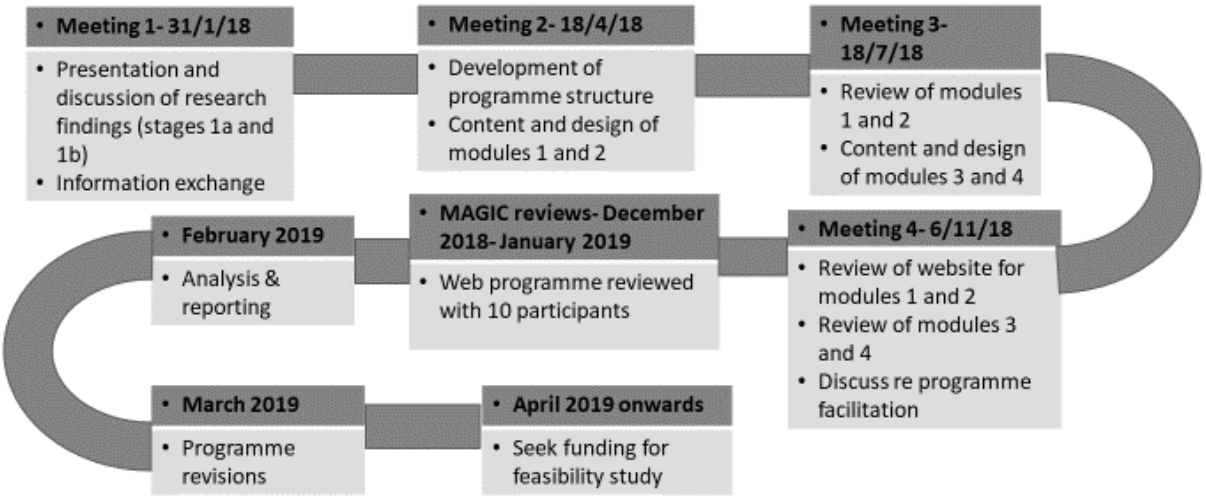


**Figure 17 Flowchart of MAGIC programme development**

The development of the MAGIC programme was an iterative process, which followed five steps (figure 18). In the first step, the findings from stages 1a and 1b and their influence on the MAGIC programme development were explored with the stakeholder

development group. These findings and their influences on the MAGIC programme development are summarised in table 25. They were classified into three components: emotional health and well-being, physical care and service issues. The practical application of these findings on the development of the MAGIC programme were considered by the group.

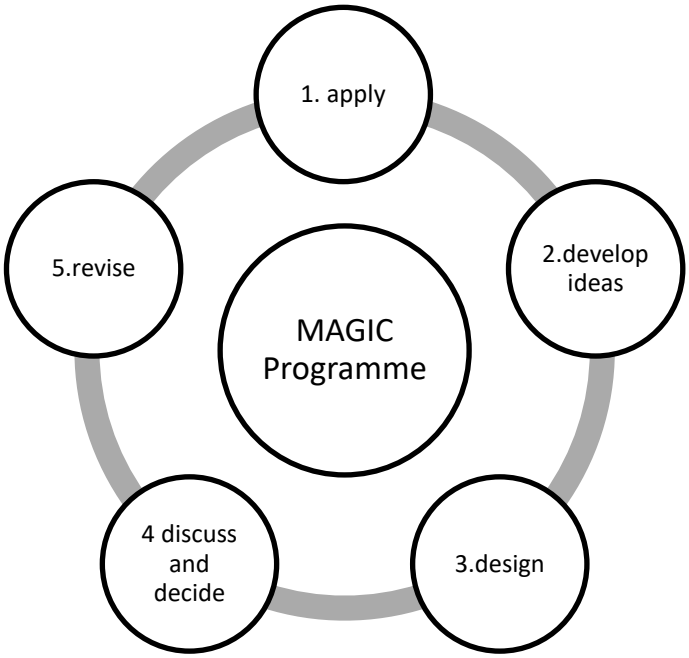


**Figure 18 Steps of MAGIC programme development**

**Table 25 Influence of findings from stage 1a and 1b on intervention design**

| Findings | Influence on intervention design |
|---|---|
| **<u>Emotional health and well-being</u>**<br><br>• ***Forming a relationship with CFD***<br>  ○ **Managing initial diagnosis CFD – negative emotions**<br><br>  ○ **Impact day-to-day life**<br>  ▪ **Change in health status (maintenance wellness)**<br>  ▪ **View CFD as part of CF**<br>  ▪ **Normalisation**<br>  ▪ **Balance vs control**<br>  ▪ **Constancy and invasiveness**<br><br>• ***Balancing CF and CFD***<br>  ○ **Unpredictability of CF**<br>  ○ **Incorporate CFD into routine CF care**<br>  ○ **Impact of CFD upon CF (knowledge deficit)**<br>  ○ **Treatment with corticosteroids**<br><br>• ***Future concerns***<br>  ○ **Dominated by CF** | Attention to both physiological and psychological issues of managing CF after the diagnosis of CFD and throughout period of adjustment<br><br>Use of peer support- videos/ vignettes of people with CFD experiences<br><br>Provision of adequate education and support to be able to respond to day-to-day variabilities and health changes<br><br>Provide education to help increase understanding of relationship between CF and CFD and why should be managed as one condition (basic physiology CFD)<br><br>Individual care planning- consider wider context of CFD, not just about achieving the numbers but achieving a sense of balance in life, realistic and achievable<br><br>Provision of adequate education and support to increase understanding of effect of corticosteroids upon CFD and appropriate management strategies |

| | |
|---|---|
| **Physical care- CFD self-management triad**<br>• ***Insulin***<br>  ○ **Uncertainties re dosing**<br>  ○ **When should insulin be taken**<br>  ○ **adherence**<br><br>• ***Blood glucose monitoring***<br>  ○ **When to test**<br>  ○ **Frequency of testing**<br>  ○ **Why test**<br><br>• ***Nutrition***<br>  ○ **Challenges/ conflicts**<br>  ○ **Role of carbohydrates**<br>  ○ **Dietary changes** | Provision of education about insulin therapy for CFD: basic physiology of insulin, insulin dosing, administering insulin<br><br>Provision of education about what, when and how to check blood glucose levels<br>Individual care planning- consider wider context of CFD, realistic goals (patient centred)<br><br>Provision of education about: food groups, meeting dietary recommendations (consideration to weight maintenance and weight improvement), carbohydrate awareness, carbohydrate counting |
| **Service issues**<br><br>• ***Insufficient knowledge***<br>  ○ **Difference CFD and type 1/ type 1 diabetes**<br>  ○ **Impact of CFD upon CF**<br>  ○ **Physiology of insulin**<br>  ○ **Insulin dose adjustment**<br>  ○ **Practical BG checking advice**<br>  ○ **Carbohydrate awareness**<br><br>• ***Lack of support*** | Provision of education will address these areas where insufficient knowledge was identified<br><br>Coordinator of the intervention will support learning and management of CFD throughout duration of intervention |

| | |
|---|---|
| ○ **How to look after CFD rather than being told what to do**<br>○ **Not focus just on numbers**<br>○ **CFD clinics implications on time**<br>○ **In adjusting insulin doses**<br>○ **Managing steroid therapy and impact on blood glucose levels**<br><br>• ***How to deliver support***<br>○ **Need for practical, realistic**<br>○ **From people with knowledge and experience CFD**<br>○ **Demonstrate insight into experience of living with CFD**<br>○ **Recognition of the individual not the disease**<br>○ **Incorporate individual into care planning**<br>○ **Role of peer support**<br><br>• ***How to provide information/ education***<br>○ **Not just telling people what to do**<br>○ **CFD clinics lack synchronicity with routine CF care**<br>○ **Manage CFD as part of CF**<br>○ **Use of Internet**<br>○ **Incorporation of short videos**<br>○ **Limited benefits of leaflets**<br>○ **Trusted source of advice** | Provision of resources to support insulin dose adjustment and steroid therapy<br><br>People with CFD to be active partners in setting goals and individual care plans<br><br>Healthcare professionals to receive training to deliver the intervention<br><br>Usefulness of peer support identified- consider setting up peer support group for people with CFD participating in the intervention (blog, face book or WhatsApp)- this will be moderated to ensure advice/support is reputable and trustworthy<br><br>Service issues outside remit of this intervention will be presented to and discussed with CF centre<br><br>Secure web-based platform to perform the staging of the intervention<br><br>Use of technology such as videos, internet-based resources<br><br>CFD will be considered as part of CF and education/support/advice will acknowledge this |

In the second step ideas for the content and design of the programme were generated. The stakeholder development group also considered what current resources were available to people with diabetes, what improvements were required to make them relevant to people with CFD and where the deficits were.

In the third step of the programme development attention was paid to the design of the MAGIC programme, this included how it would be structured and what it would look like. Considerations to the use and format of videos, personal experience vignettes and the use of pre-existing resources were made. A prototype of the website was reviewed by the stakeholder development group. People with CFD who were happy to share their experiences with others either in the form of videos or vignettes were recruited from the centre's CFD clinic and from within the stakeholder development group. Ten people with CFD were approached when they attended CFD clinics to explore the possibility of them assisting with personal experience videos and vignettes. The project was explained to them along with the purpose of the videos/ vignettes. A follow up phone call was made a week to ten days later to verify if they were happy to help. Six people were happy to assist with vignettes, two with videos. A member of the stakeholder group also volunteered to assist with videos. Those who helped with vignettes gave written permission for their anonymised quotes to be used in the MAGIC programme. Individuals who helped with videos gave written permission for their self-recorded videos to be used in the MAGIC programme.

The fourth step involved the stakeholder development group working through the programme content to check language, illustrations, appropriateness of videos, use of personal experience vignettes/videos and identified resources.

The fifth stage of the development of the MAGIC programme involved the stakeholder development group revising and making any necessary amendments to the programme.

The MAGIC programme was developed in partnership with the digital content manager. A close working relationship between the project lead and the digital content manager was established, he also attended the stakeholder development

group meetings to develop ideas and gain understanding of the practical requirements of the programme. The platform was developed using HTML5, CSS and JavaScript technology for the front end (visible parts of the website) and PHP5, PHPMYADMIN for the back end (infrastructure and data bases). The MAGIC programme is hosted outside the NHS network but runs on an encrypted certified hosting service. The connection between the hosting and the client is secured by Secure Sockets Layer (SSL). When a user enters the SSL-protected Platform, the SSL certificate automatically creates a secure, encrypted connection. The SSL certificate uses a SHA-2 digest method and 2048-bit encryption to protect sensitive data. The whole platform and its database are backed up daily on the hosting solution. This will secure the content and data in case of data loss. The platform is password protected and can only be accessed by a given username and password. Users will need to be pre-registered to obtain a personal login to access the content of the modules.

## 5.2 Results

### 5.2.1 The stakeholder development group meetings

In line with the recommendations of stage 1a the stakeholder development group discussed the use of the term CFD. The decision was made to use the term CFD for this project; it was believed that this highlighted the greater causal relationship between CF and diabetes and promoted more harmonious management. Throughout the development of the MAGIC programme the stakeholder development group remained mindful of its focus on 'managing abnormal glucose in cystic fibrosis.' To remain true to this, and taking the timeframe into consideration, the group decided to focus the programme on the knowledge and skill acquisition required to self-manage CFD. This was identified as the CFD self-management triad (insulin, blood glucose, nutrition) and 'what is CFD?' It was decided that the MAGIC programme should have a basic nuts and bolts module, which was aimed at people new to CFD, progressing to more advanced modules for advanced learning.

The four stakeholder development group meetings lasted from 10.00 to 15.00 with a scheduled break for lunch. To help maintain focus throughout the meetings group

members were asked, at regular intervals throughout the day, if they required a break. Additional comfort and refreshments breaks were then held as and when required.

As part of introductions in the first meeting the co-design process was discussed - this included equality. It was discussed with the stakeholders that people with CFD and healthcare professionals were considered equal and that everyone had a say in the decision-making processes. At least one member of my academic team attended each stakeholder meeting. They were not an expert in CFD or directly involved in designing and producing the programme and were therefore able to be non-biased. They were able to provide support throughout the meetings by seeking clarification, stimulating discussion and challenging ideas. To try and ensure that all group members had the opportunity to comment or contribute to the development of the MAGIC programme I would ask for additional comments and encourage involvement from all group members at regular intervals throughout the meetings. A link person was appointed for each meeting. Their role was to maintain contact with the stakeholders at home and monitor for questions that may have occurred via the chat feature of BlueJeans and to identify any technical difficulties encountered. The stakeholder group meetings were relaxed and informal to facilitate discussion and put members at ease. Content including text, videos and illustrations for the MAGIC programme was also emailed to group members between the meetings giving further opportunity for comments and input. This was particularly useful in areas that we struggled with during the meeting. For example, there was a difference of opinions between medical professionals in the group and people with CFD with regards to sick days rules and how to make insulin adjustments. People with CFD didn't want to have to have to do calculations when unwell they wanted simple rules on what to do with their insulin whereas some of the medical professional felt we should be giving patients the tools to make decisions rather than tell them what to do. After discussing responses and experiences the stakeholder group made the decision to follow the view of people with CFD as they have firsthand experience of such situations and how they would like to manage them. The MAGIC programme was developed through a shared decision-making process, by establishing a close working relationship through the sharing of knowledge and

expertise between people with CFD and healthcare professionals caring for people with CFD.

The first meeting generated the topics, considered the structure and discussed the theoretical underpinnings of the MAGIC programme. Figures 19 and 20 were developed after the meeting, they summarised the ideas for the content and structure of the MAGIC programme. The content for modules one and two was then written by the project lead between the first and second meeting.

In the second meeting the stakeholder development group reviewed the proposed ideas and modules for the MAGIC programme and its structure, making modifications where it was deemed appropriate; this included ensuring the first module contained some basic information on hypoglycaemia management. The content of the first two modules was then reviewed, paying particular attention to readability and use of language. The preference for short, clear sentences which avoided the use of too many abbreviations was deemed preferential. Attention was also paid to the appropriate use of illustrations and videos. This included making sure all images were clear, of the appropriate size and easy to understand. Existing videos linked to the MAGIC programme, via their websites, were reviewed for their appropriateness to people with CFD, sound quality and educational content. These included: 'what is diabetes video?' produced by Diabetes UK and a series of carbohydrate counting videos produced by Norfolk and Norwich University Hospital. All videos made by the stakeholder development group were checked for sound quality, clarity and comprehension.

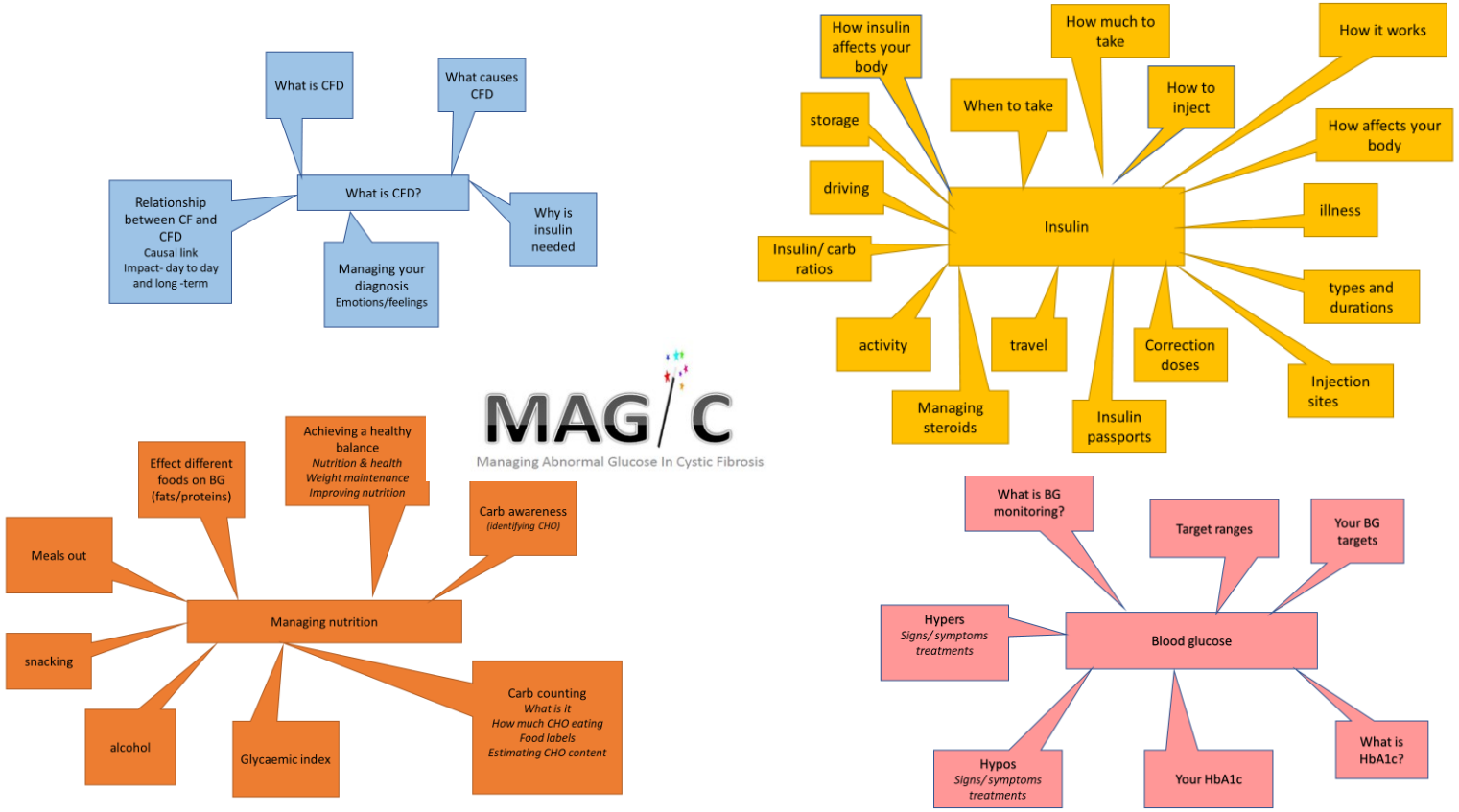


**Figure 19 Ideas for the MAGIC Programme**

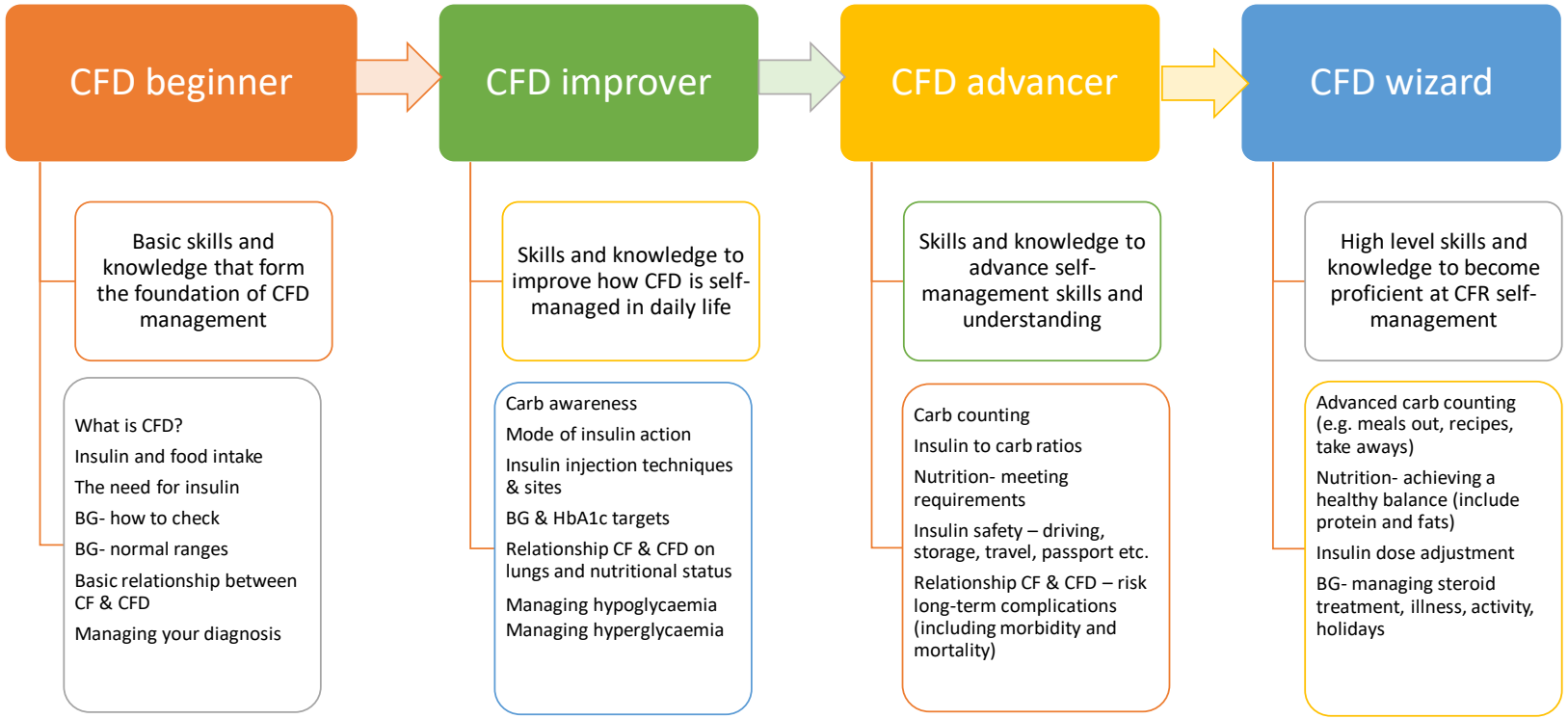


**Figure 20 Ideas for the modules of the MAGIC programme**

In the third meeting the group viewed and discussed the prototype of the web-based programme for the CFD-beginner module, which had been developed between the second and third meeting. This provided greater visual clarity of the MAGIC programme. The content of the final two modules were then reviewed.

The stakeholder development group believed that personal experiences would add to the quality of the MAGIC programme. They were believed to be a beneficial part of the learning process because people with CF are recommended to not have face-to-face contact with each other and hence have limited opportunities to share experiences. The stakeholder development group explored how personal experiences of people with CFD could be incorporated into the programme. Some example questions for exploration were drafted to try out with volunteers who were happy to record videos. The decision was made to produce some self-recorded 'vlog style' videos and some written quotes reflecting personal experiences of living with and managing CFD.

In the final stakeholder development group meeting the group viewed and discussed the web-based programmes developed for the CFD beginner and CFD improver modules, this included: overall structure, content, a demo personal experience video, personal experience quotes, the formatting of the test your learning sections and the interactive components. Feedback from the group then guided the development of the web-based format for the final two modules.

The stakeholder development group also briefly explored the role of the facilitator within the MAGIC programme to help identify the frequency and type of support they would provide. Time points for facilitation have been highlighted in the programme curriculum, see appendix 19 for the example curriculum of CFD beginner. The role of the facilitator was clarified in terms of the supporting role they would have with patients undergoing the programme. Their role would be in facilitating education and acquisition of skills and not the provision of clinical support. It has not been confirmed how the support from the facilitator will be best provided and this will be developed in the next stage of research.

## 5.2.2 The MAGIC programme

### 5.2.2.1 Overall design

The four modules that made up the MAGIC programme are described in more detail below, with illustrative examples of content and the application of the theoretical underpinnings.

The MAGIC programme was designed as a patient focussed, staged approach to learning; with the aim to develop self-efficacy, knowledge and skills as the participant progressed through the modules. All the modules contained a mixture of text, educational videos, illustrations, interactive tasks, personal experience quotes/ videos and a 'test your learning' quiz. Three educational videos were produced by the stakeholder development group. These were: what is CFD? how to use a blood glucose monitor and how to give an insulin injection (figure 21). These videos were less than two minutes long and included both subtitles and narration. The 'what is diabetes video' by Diabetes UK and series of carbohydrate counting videos by Norfolk and Norwich University Hospital were reviewed. The stakeholder development group decided these videos were well made and applicable to the educational needs of people with CFD.

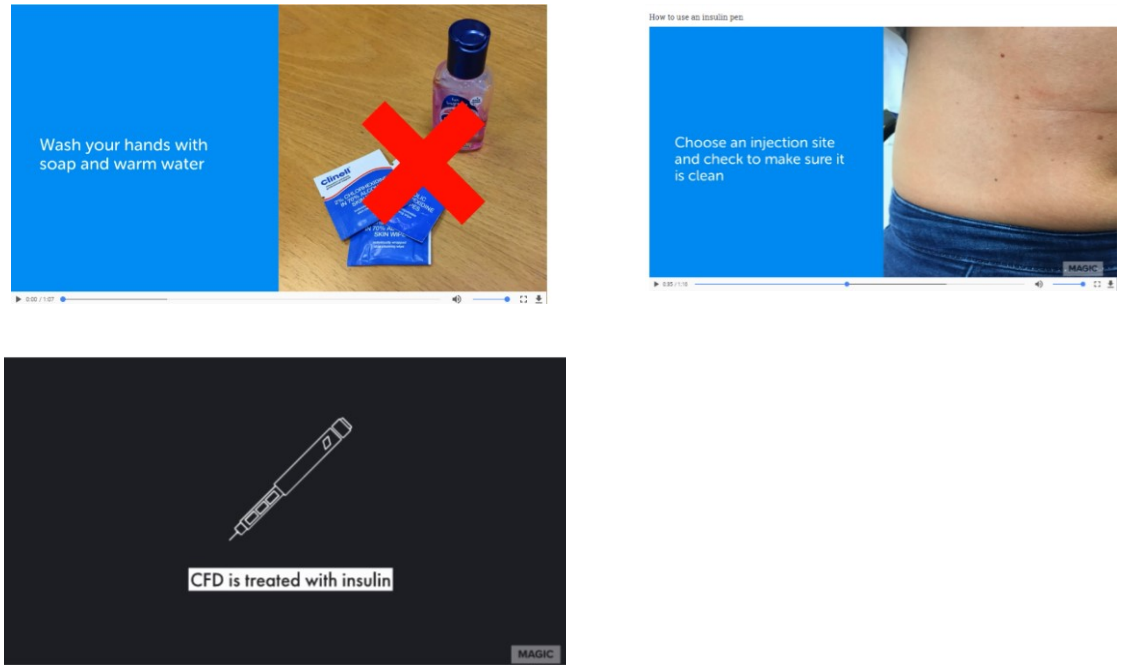


**Figure 21 Examples of educational videos produced by the MAGIC programme**

The stakeholder development group have not specified that someone with CFD should complete all four modules of the programme; it is dependent upon the individual and their needs. It would be the role of the programme facilitator to work with the individual and establish their specific requirements. As the programme takes a staged approach to learning it is anticipated that most individuals will work through the modules from the CFD beginner to the CFD wizard. However, an individual may choose not to do this and perhaps dip in and out of modules choosing areas of interest to them instead. A homepage introduces the participants to the MAGIC programme (figure 22).

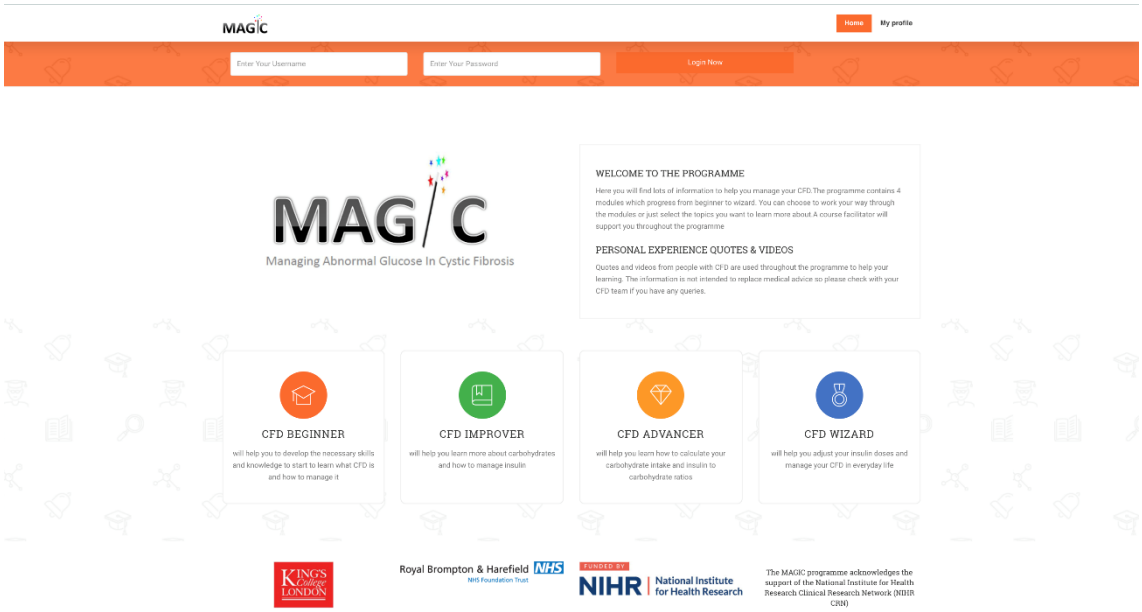


**Figure 22 Screenshot- homepage**


**Figure 23 Screenshot CFD beginner**

CFD beginner (figure 23), the first module, was designed for someone new to CFD with the assumption being they had very little knowledge or experience of CFD. The stakeholder development group considered it as the foundation to CFD management. It focussed on basic dietary, insulin and blood glucose management- the minimum knowledge someone with CFD was expected to have. The stakeholder development group decided to include some basic information about hypoglycaemia in this module, as a safety net. Therefore, illustrations of the symptoms of hypoglycaemia (figure 24) and the hypoglycaemia flowchart (figure 25), developed for the CFD improver module, were also included here.

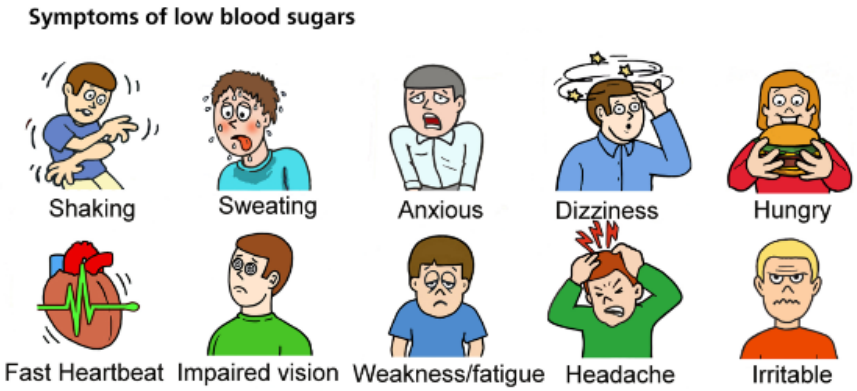


**Figure 24 Illustration of hypoglycaemia symptoms**

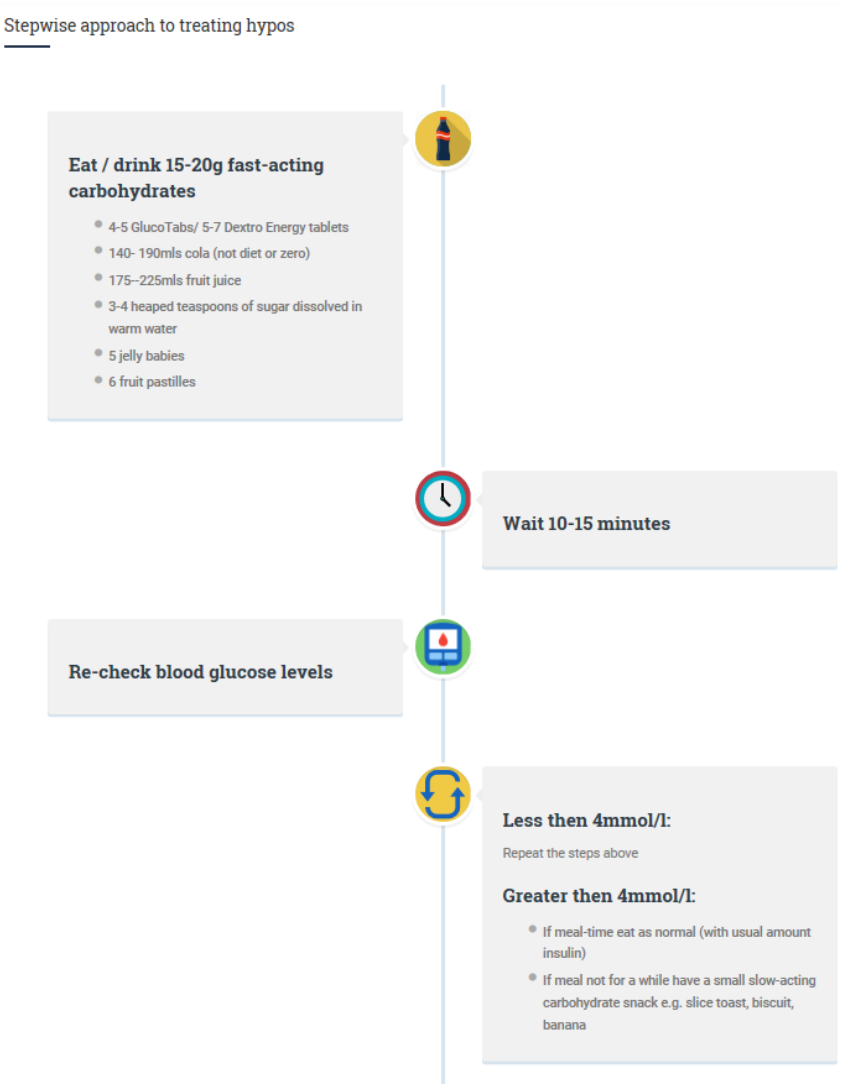


**Figure 25 Hypoglycaemia flowchart**

CFD improver (figure 26), the second module, built upon the learning from CFD beginner with increasing levels of depth and complexity. This included a focus on increasing knowledge about different types of carbohydrates and their effects, insulin administration and mode of action.

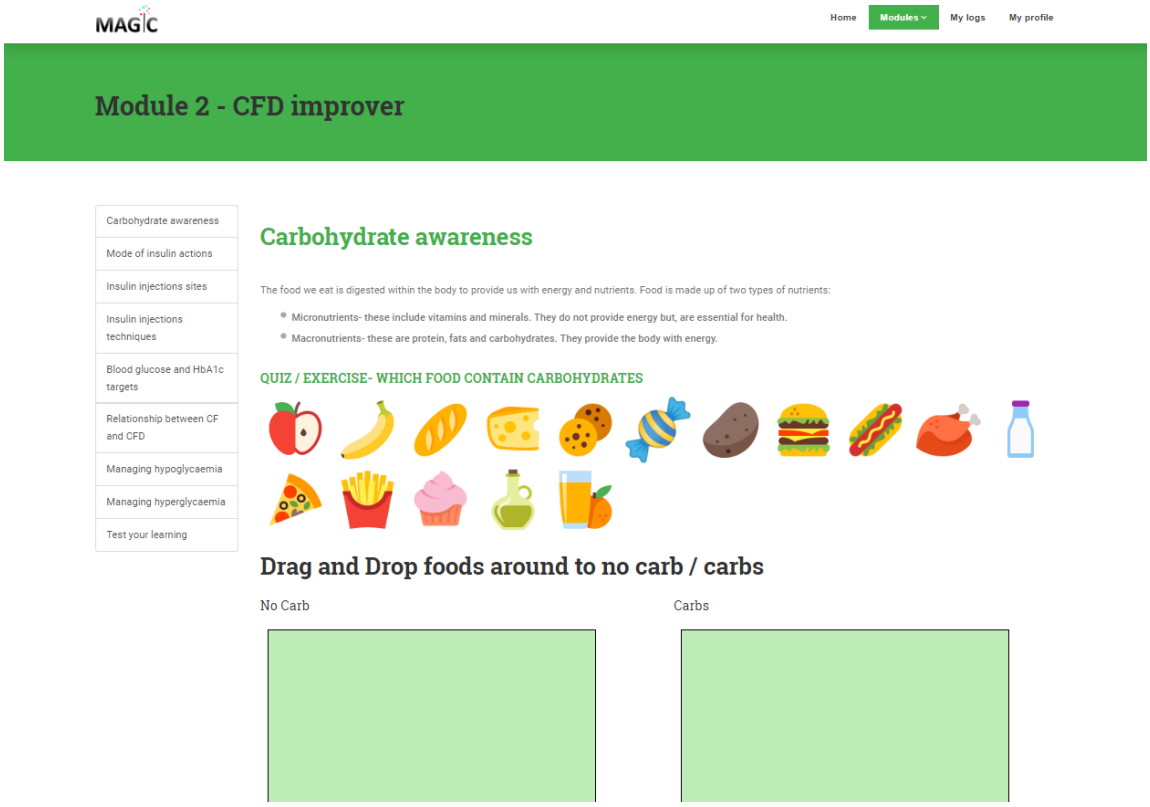


**Figure 26 Screenshot CFD improver**

The third module, CFD advancer (figure 27), aimed to develop more advanced skills in CFD management which included carbohydrate counting and calculating insulin to carbohydrate ratios. These are more complex skills to learn and develop and therefore more practical examples and illustrations were included to help facilitate learning in this module.

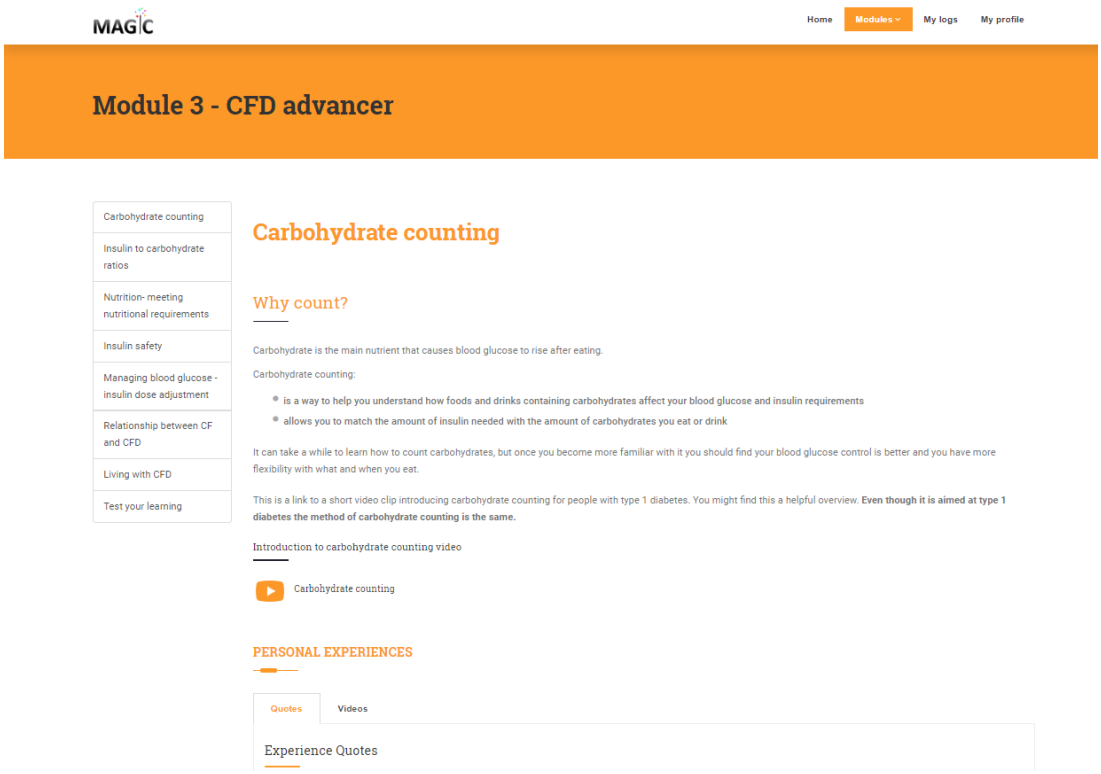


**Figure 27 Screenshot CFD advancer**

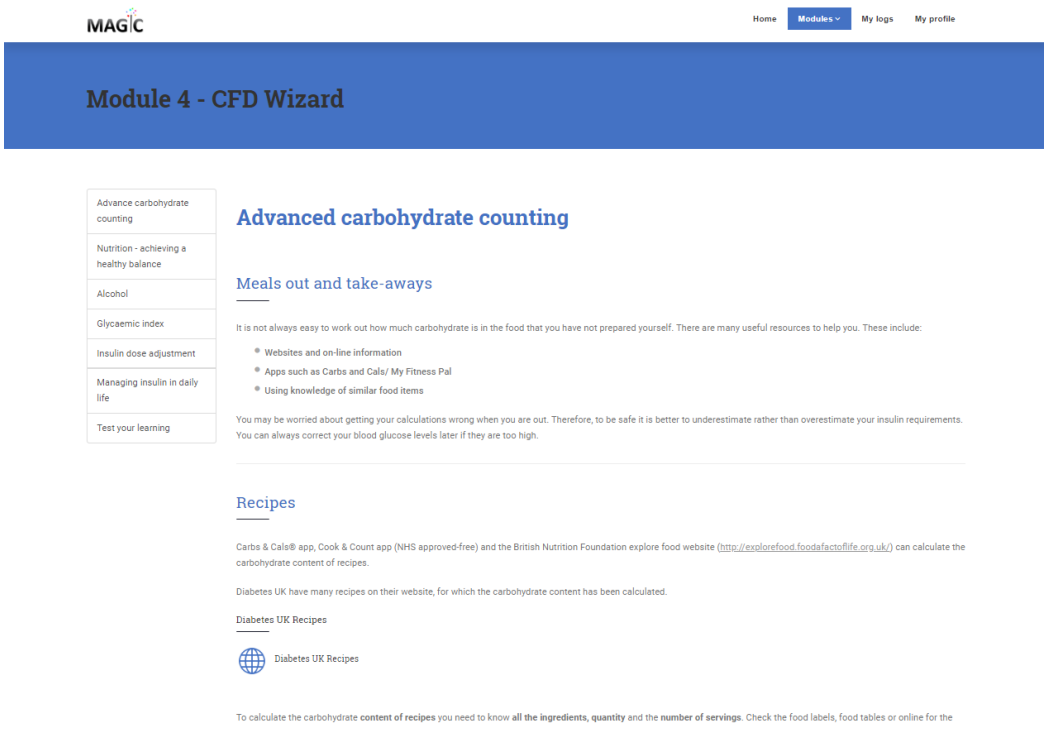


**Figure 28 Screenshot CFD wizard**

The final module- CFD wizard (figure 28) addressed more complex CFD management skills which included: insulin dose adjustment and managing insulin in daily life (eating out, travelling, alcohol, illness and exercise).

#### 5.2.2.2 Adult learning styles

The four adult learning styles: visual, aural, read/write and kinaesthetic (Fleming 2001) were carefully considered when designing the MAGIC programme (table 26). Figure 29-32 are visual representations of how the MAGIC programme applied adult learning styles.

**Table 26 Application of adult learning styles to the MAGIC programme**

| Learning style | MAGIC programme examples |
|---|---|
| **Visual** | Graphs<br>Charts<br>Illustrations<br>Tables<br>use of colour<br>Personal experience videos<br>Educational videos |
| **Aural** | Personal experience videos<br>Educational videos<br>Discussion with the facilitator |
| **Read/write** | Text- use of short sentences, highlighting key points,<br>Personal experience quotes<br>Personal logs<br>Completing food and blood glucose diaries<br>Following instructions |
| **Kinaesthetic** | Practical activities<br>Completing test your learning quiz<br>Carrying out example scenarios/activities<br>Carrying out tasks in real life |

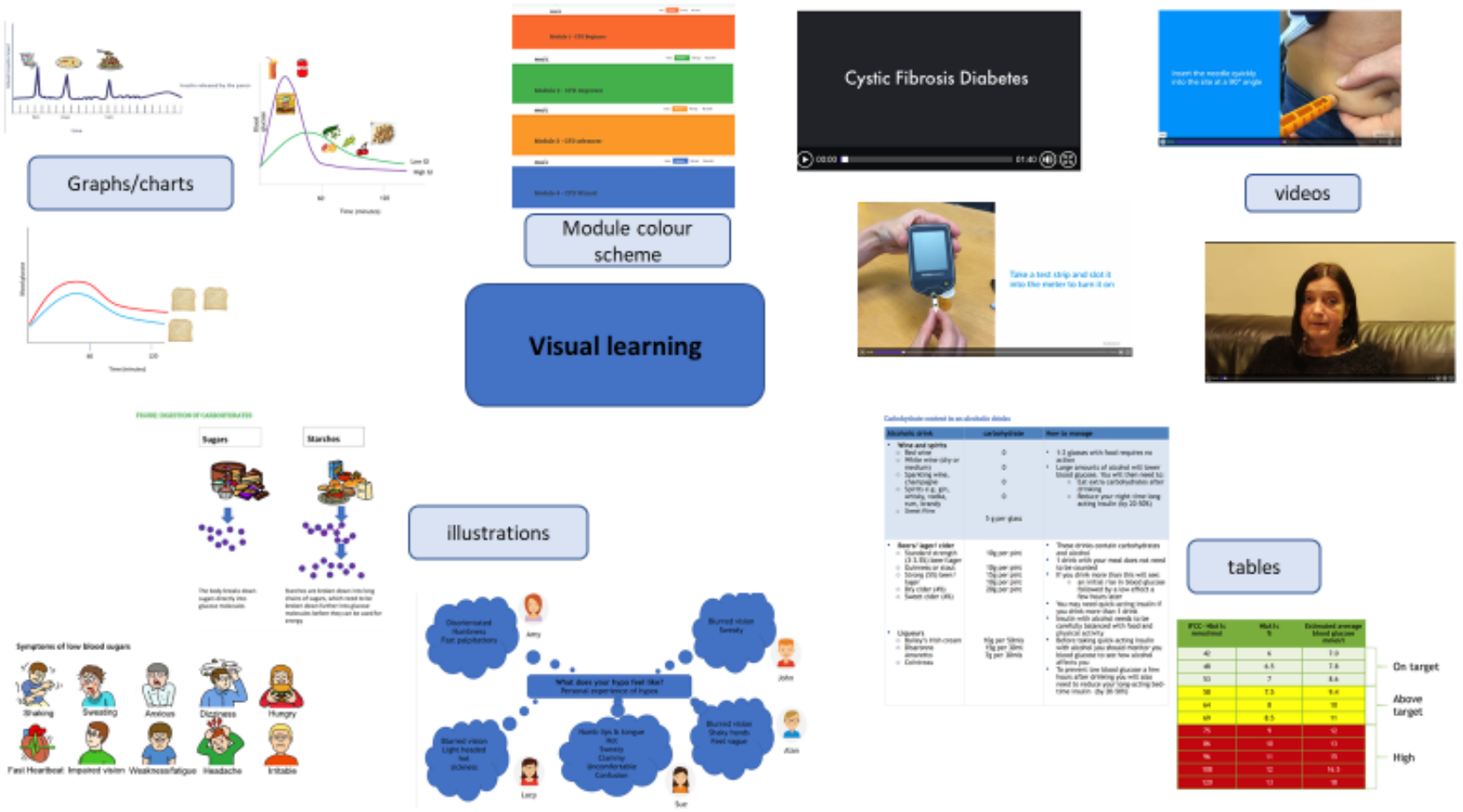


**Figure 29 Examples of visual learning styles in the MAGIC programme**

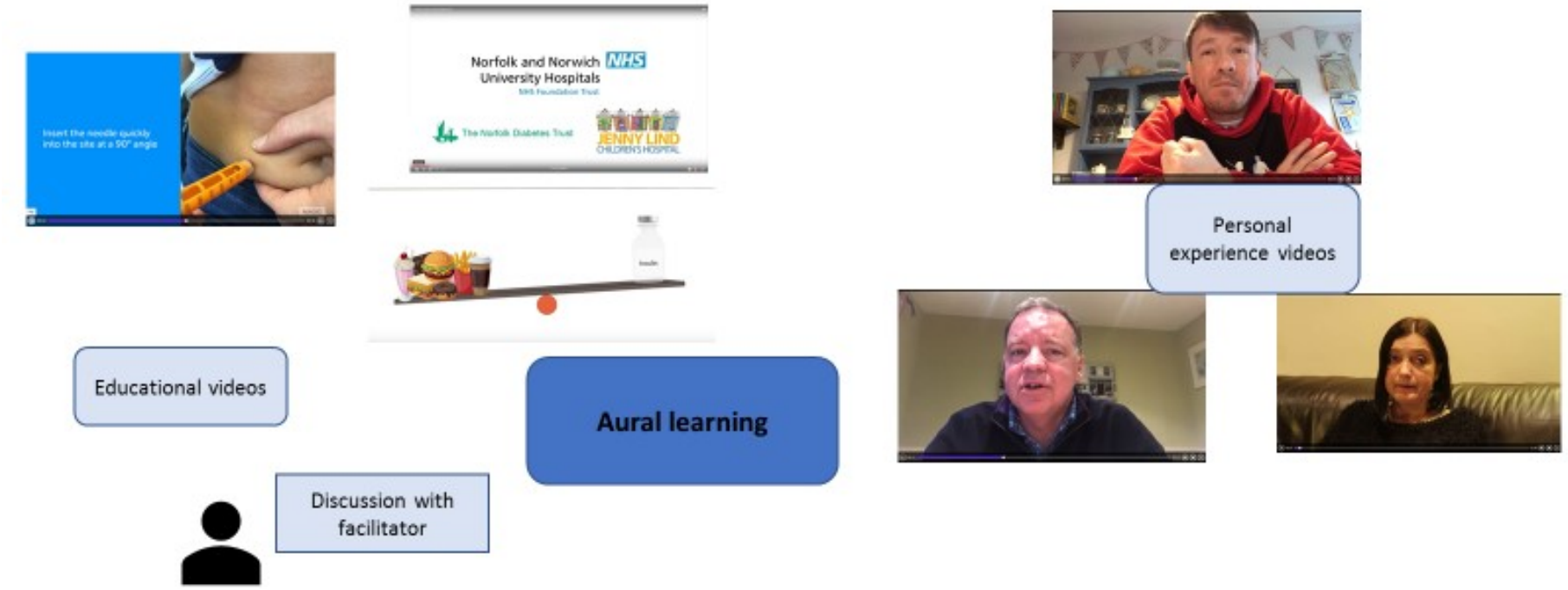


**Figure 30 Examples of aural learning styles in the MAGIC programme**

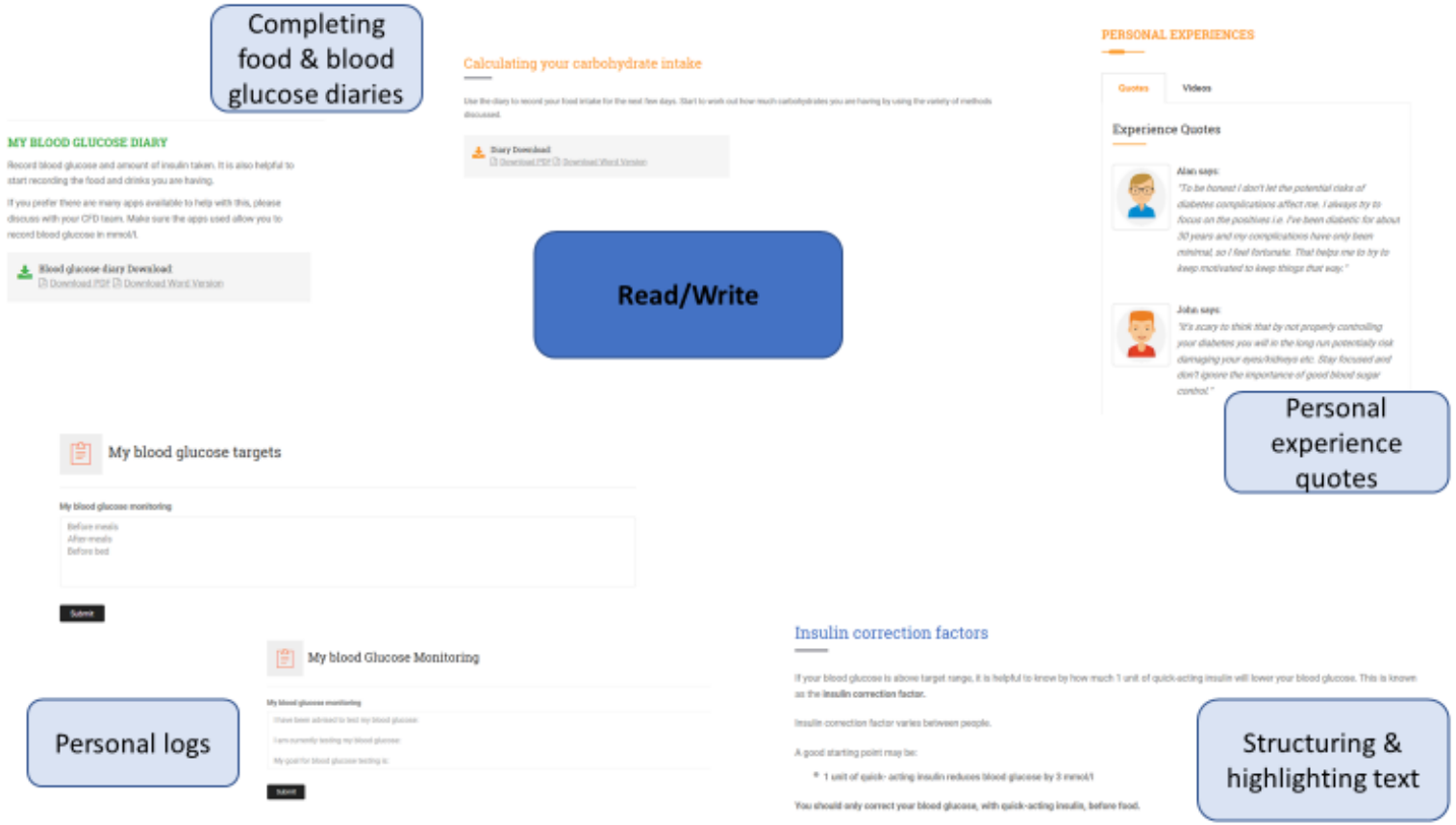


**Figure 31 Example of read/write learning styles in the MAGIC programme**

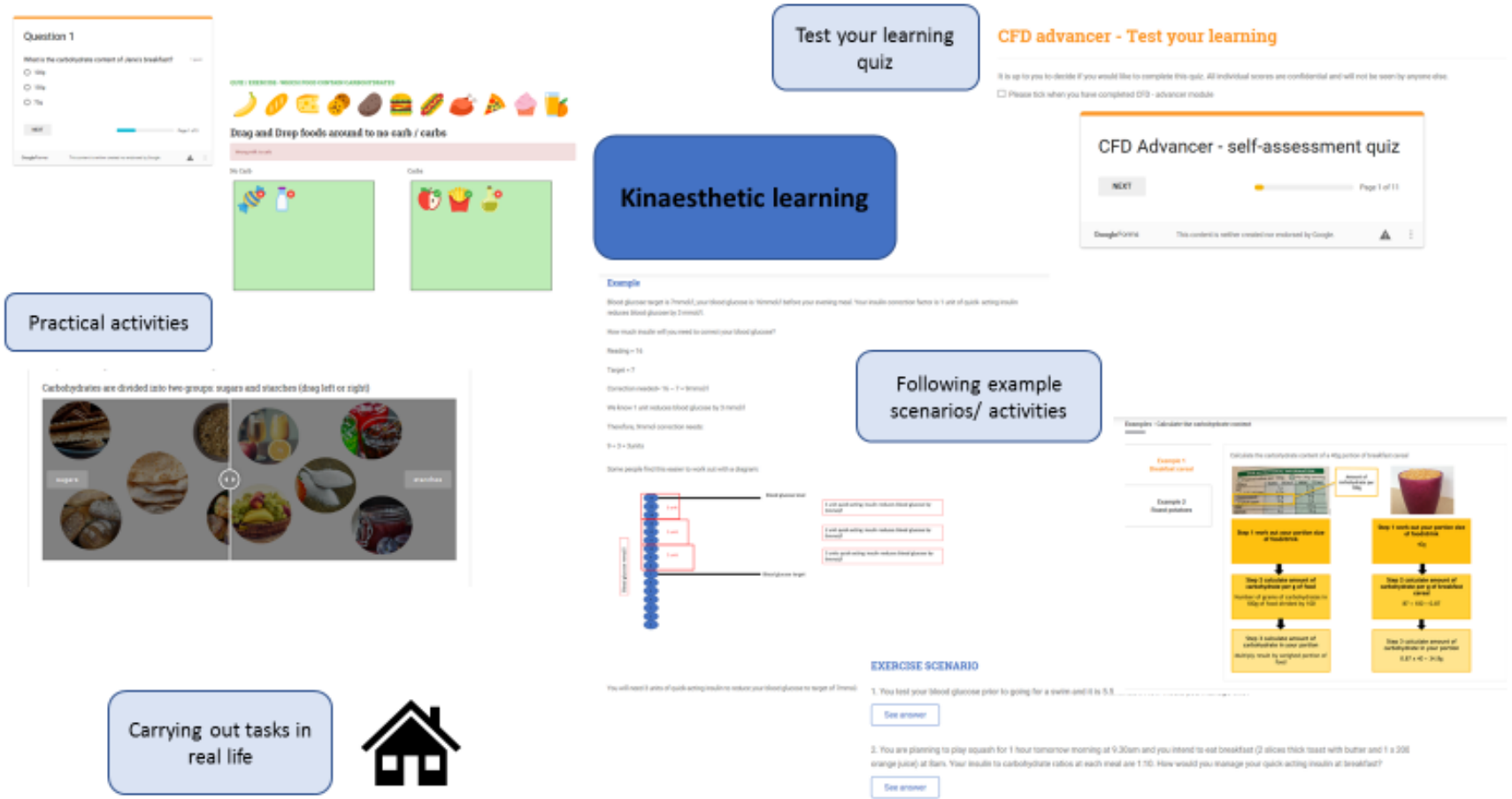


**Figure 32 Examples of kinaesthetic learning styles in the MAGIC programme**

### 5.2.2.3 Self-management framework of chronic illness

As stated in chapter two the self-management framework of chronic illness (Corbin & Strauss 1988) recognises three important components to self-management of chronic illness namely: medical management, role management and emotional management. All three of these roles have been addressed within the MAGIC programme (table 27).

**Table 27 Mapping of module content and self-management roles**

| | CFD beginner | CFD improver | CFD advance | CFD wizard |
|---|---|---|---|---|
| **Self-management role** | | | | |
| **Medical management** | What is CFD? | Carb awareness | Carbohydrate counting | Advanced carbohydrate counting |
| | Why does CF cause diabetes? | Mode of insulin action | Insulin to carbohydrate ratios | Nutrition- achieving a healthy balance |
| | Why do we need insulin? | Insulin injection sites | Nutrition- meeting nutritional requirements | alcohol |
| | What happens without insulin? | Insulin injection techniques | Insulin safety | Glycaemic index |
| | Relationship between CF & CFD | Blood glucose & HbA1c targets | Blood glucose-insulin dose adjustment | Insulin dose adjustment |
| | Symptoms of CFD | Relationship between CF & CFD | Relationship between CF & CFD | |
| | How is CFD treated? | Managing hypoglycaemia | | |
| | Blood glucose | Managing hyperglycaemia | | |
| | Managing hypoglycaemia | | | |
| **Emotional management** | Beginning to live with CFD | | Living with CFD | |
| **Role management** | | | | Managing insulin in daily life |

### 5.2.2.4 Self-efficacy theory

The four main influences on self-efficacy are mastery experience, vicarious experiences, verbal encouragement and psychological state (Bandura 1977b); these have been addressed within the MAGIC programme. Mastery experience builds upon knowledge, skills and experience through a staged approach to learning. Figure 33

highlight how the complexity of the modules, and hence the associated skills and knowledge required, increased from CFD beginner module to the CFD wizard module. Mastery experience is also acquired through the direct experience of successfully mastering a task. There are many tasks involved in the MAGIC programme to manage CFD, including: giving an insulin injection correctly, calculating the carbohydrate content of food, managing blood glucose effectively during illness and treating hypoglycaemia successfully. The identification and competition of tasks will vary according to the individualised requirements of the participant and will be driven by their learning, experience and skills requirements.

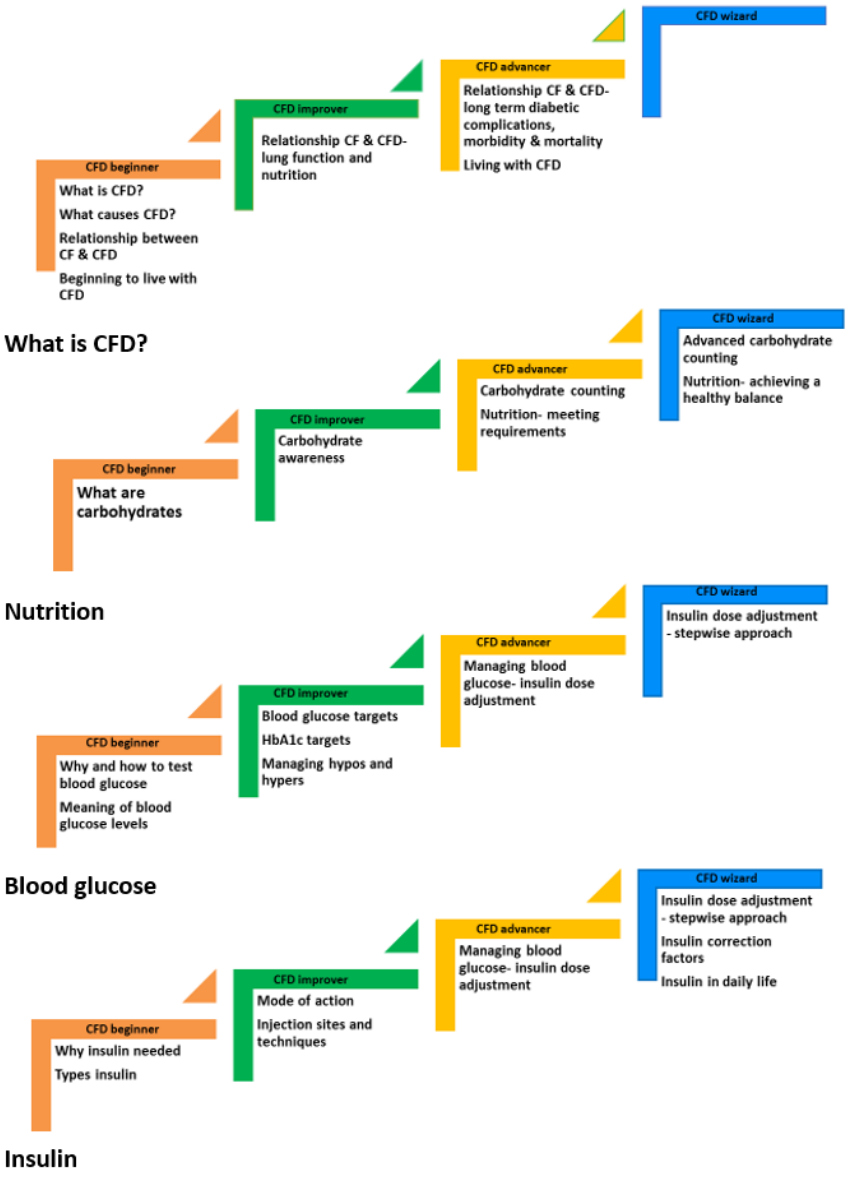


**Figure 33 Mastery experience**

Vicarious experiences involve learning from others and includes the observations of behaviours and actions. Due to cross infection risks the MAGIC programme will be delivered on a 1:1 basis via the Internet. The opportunity to learn from others in this context is more limited, however the inclusion of personal experience videos gives the participants opportunities to listen to others with CFD talk about their experiences and provides a vehicle for learning.

Verbal encouragement influences an individual's belief in their ability to self-manage and acknowledges that many challenges and difficulties may occur. The MAGIC programme facilitator will provide support while participants complete the MAGIC programme. Some messages of persuasion, encouragement and acknowledgement of difficulties are also present in the personal experience videos. The MAGIC programme offers messages of normalistaion throughout. These include the acknowledgement that carbohydrate counting and insulin to carbohydrate ratios are not always easy concepts to understand, but they get easier with time. The living with CFD sections in CFD beginner and CFD advancer modules helps to normalise that living with CFD is challenging and can impact upon feelings; these sections include personal experiences quotes and videos to help deliver these messages (figure 34).

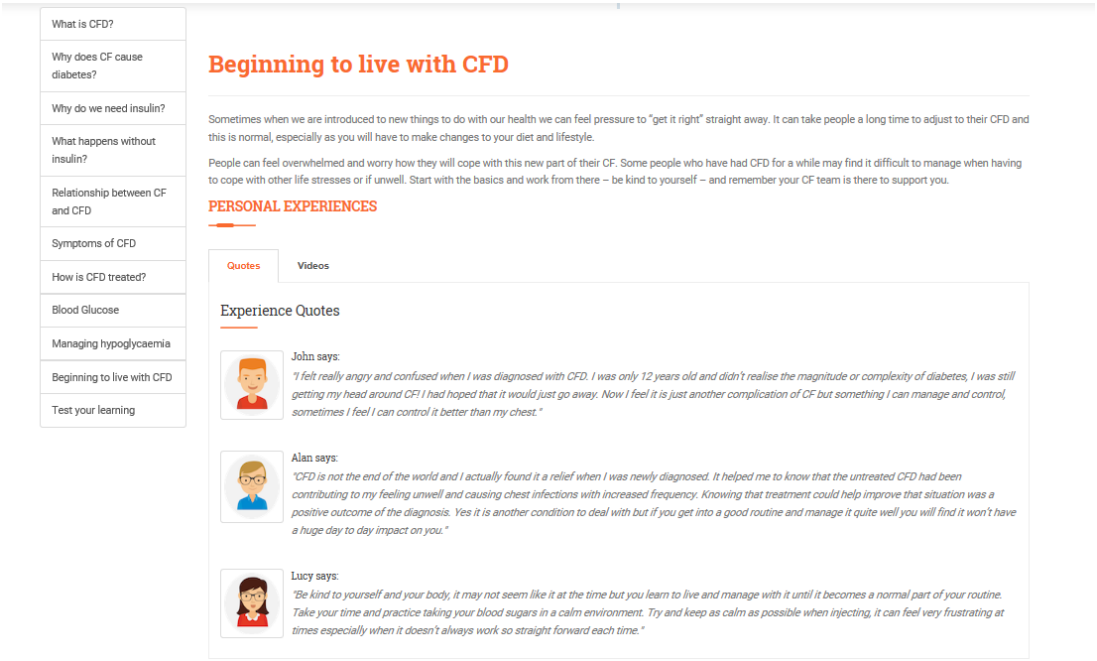


**Figure 34 Beginning to live with CFD from CFD beginner module**

The main focus of the MAGIC programme was on self-management skills and the knowledge required to manage abnormal glucose in CF. Therefore, addressing psychological challenges such as mood, anxiety or stress were not addressed in detail within the MAGIC programme. The sections on living with CFD addressed some psychological and emotional challenges and included recommended resources and suggestions on how to seek further support and advice. The use of personal experience quotes and videos, from people living with CFD, provided insight into some of the challenges faced and how these were managed.

#### 5.2.2.5 NICE criteria

The MAGIC programme was developed to meet the NICE criteria (NICE 2016). Table 28 demonstrates how NICE criteria were considered when designing the MAGIC programme. A curriculum was developed to support each module of the MAGIC programme, it addressed aims, learning outcomes, content and resources. An example of a page from the module curriculum for CFD beginner is illustrated in figure 35, the full CFD beginner curriculum can be found in appendix 19.

**Table 28 How the MAGIC programme meets NICE criteria**

| NICE criteria | Examples from the MAGIC programme |
|---|---|
| • **Be evidence-based** | Qualitative finding from stages 1a and 1b<br>Use of expert healthcare professional and people with CFD in co-designing the programme<br>Use of evidence-based recommendations e.g. FIT, UK Gov. |
| • **Have specific aims and learning objectives and support the development of self-management** | Aims and learning outcomes specified for the main topics of each module (example in figure 35). |
| • **Follow a structured written curriculum driven by theory** | Curriculum developed for each module. Informed by: self-efficacy theory, self-management framework and adult learning styles |
| • **Be delivered by trained facilitators**<br>• **Be quality assured**<br>• **Have its outcomes audited regularly** | Outside the scope of this study. |

| CFD BEGINNER<br>Learning goals | content | Resources/ notes |
|---|---|---|
| • Be able to describe what CFD is | **What is CFD?**<br>• Brief explanation diabetes caused by lack of insulin and resultant increase in blood glucose<br>• Discussion of delay and reduction in insulin-<br>• Who affects | • video – brief overview CFD<br>• module text |
| • Know where insulin is produced<br>• Know why cells need glucose<br>• Understand how insulin lowers blood glucose levels<br>• Understand that normally insulin is produced automatically | **Why do we need insulin?**<br>• Explanation of journey from carbohydrates in food we eat to energy for use by our bodies.<br>• Discussion what happens in someone without diabetes | • Module text<br>• Diagram food to energy<br>• Chart normal insulin production |
| • Discuss what happens when the body does not produce enough insulin | **What happens without insulin?**<br>• Discussion of consequences of not having enough insulin- increase glucose production by liver, breakdown of body's protein and fat stores and consequent weight loss, muscle weakness, tiredness. Spill over of glucose into urine and subsequent polyuria and polydipsia. | • Module text<br>• Diabetes UK what is diabetes video https://www.diabetes.org.uk/diabetes-the-basics/diabetes-and-the-body |
| • Identify why having CF causes CFD<br>• Be aware that CFD is not preventable | **Why does CF cause diabetes?**<br>• Discussion of link between pancreatic disease in CF and development of CFD | • Module text |
| • Be aware of the effects of CFD on CF | **Relationship between CF and CFD** | • Module text |

**Figure 35 Example of aims and learning outcomes as identified in the curriculum**

## 5.3 Method - Stage 2b review of MAGIC programme

### 5.3.1 Research design

The aim of this stage was to determine the face validity of the MAGIC programme with people who had not been previously involved in any part of the study. Cognitive interviewing techniques were used to focus on participants' experiences and understanding of the MAGIC programme and its content.

### 5.3.2 Research setting

All participants received their care at a Regional Adult CF centre. All interviews took place in a private room, either within the research facility or on the CF ward, at this CF centre.

### 5.3.3 Participants

It was originally decided that the MAGIC programme was going to be reviewed with people who had CFD, however the early modules are designed for people new to CFD

and the decision was therefore made to also include people without CFD as the reviewers of the first two MAGIC programme modules. Due to time constraints and the development phase taking longer than expected there was a limited widow of opportunity for recruitment. Participants were initially identified from outpatient clinic schedules and ward lists. The participants were purposively sampled to meet the inclusion criteria of having CF and/or CFD and being over 16 years of age. Twenty-one letters were sent out in stages. The aim was to recruit 8-10 people. Eleven people agreed to take part in the study however one withdrew prior to the interview due to ill health. Study visits coincided with outpatient clinic appointments or ward admissions. This reduced requirement for additional hospital attendance. Interviews took place between December 2018 and January 2019. No participant had taken part in any previous MAGIC study related activity. Participants received a £20 'thankyou' voucher for taking part in the study, and if required travel expenses were reimbursed.

### 5.3.4 Ethical considerations

Prior to commencing the study, approval was received from: London-South East Research Ethics committee- 17/LO/0377 (appendix 7), NHS Health Research Authority (appendix 8) and Royal Brompton and Harefield NHS Foundation Trust (appendix 9). Informed written consent, including agreement for digital audio-recording and note taking was obtained prior to starting the interviews. A copy of the participant's consent form was stored in their electronic patient record. The audio recordings were stored on a password protected computer.

### 5.3.5 Data collection

Each participant reviewed one module with the researcher. A range of cognitive interviewing techniques including: exposure to test material, think aloud, observation, probing and task completion were used to verify understanding of content and instructions, identify gaps and appraise the visual appeal of the programme (D'Ardenne et al. 2015). Table 29 illustrates how cognitive interviewing techniques were applied to the MAGIC programme review process. The interviews were digitally recorded, and notes were taken throughout the process.

**Table 29 Application of cognitive interviewing techniques in the MAGIC programme review**

| Cognitive interviewing technique | Example |
|---|---|
| **Exposure to test material** | Participants were asked to work their way through the module, trying tasks, navigating around the website, watching videos and completing quizzes |
| **Think aloud** | Participants were asked to talk through their thought processes when completing tasks/ activities |
| **Observation** | Participants were observed navigating around the module and its content |
| **Probing** | Participants were asked questions to review their understanding for example from reading that section can you tell me what carbohydrates are? What do you think that section is trying to tell you? Was there anything in that section wasn't clear? Was there anything missing? What could be done to make this clearer? |
| **Task completion** | Participants were observed carrying out tasks such as the drop and drag activities, calculating the carbohydrate content of meal activity, working out insulin correct factors and test your learning quiz. Questions were used to clarify their understanding of instructions and processes |

### 5.3.6 Data analysis

After each review the interview notes were written up; where there was ambiguity the interview recordings were listened to for further clarity. The findings were given classification titles (table 30) and action points were identified.

**Table 30 Classification of findings**

| classification | description |
|---|---|
| **omission** | Missing information- words/ letters/ punctuation |
| **error** | Information/ objects in the wrong place<br>Information not correct |
| **visual** | Any issues with use of colour, illustrations, diagrams, highlighting text |
| **comprehension** | Difficulty understanding wording/ illustrations/ graphics/ videos<br>Use of language |
| **technical** | Anything involving sound, videos, moving images or links to websites |
| **other** | Anything else |

## 5.4 Results

Ten people participated in the interviews, these lasted between 71 and 134 minutes (median 107 minutes). Demographic information for the participants is shown in table 31. Six of the study participants had CFD, three did not have CFD and one had impaired glucose tolerance, for which he was not currently receiving treatment. Each module was reviewed by at least two participants. An example of review findings is shown in figure 36; it summarises the findings, highlights action points and applies classifications.

**Table 31 Demographics of study participants**

| participant | gender | Age (years) | CFD status | Duration of CFD | Module reviewed |
|---|---|---|---|---|---|
| **1** | M | 36 | IGT- not on treatment | N/A | CFD beginner |
| **2** | M | 42 | CF | N/A | CFD beginner |
| **3** | M | 48 | CFD | > 5 years | CFD improver |
| **4** | M | 43 | CF | N/A | CFD improver |
| **5** | M | 33 | CF | N/A | CFD improver |
| **6** | M | 44 | CFD | >5 years | CFD advancer |
| **7** | M | 56 | CFD | >5 years | CFD advancer |
| **8** | F | 32 | CFD | >5 years | CFD advancer |
| **9** | F | 43 | CFD | >5 years | CFD wizard |
| **10** | F | 45 | CFD | >5 years | CFD wizard |

MAGIC review – participant 4 13th December 2018 (CFD Improver)

| Topic | Discussion | Action | Classification |
|---|---|---|---|
| **Carbohydrate awareness** | Drag and drop- didn't notice well done when it appeared, perhaps a tick or cross instead of well-done, also these could remain next to the pictures as struggled to remember which was right or wrong<br>Potato-not sure it was a potato<br>Milk- not sure of image<br>Oil- didn't know what this was<br>Candy- should say sweets- label is wrong it is a carb<br>Big area for drag and dropping | Review how answers are highlighted right or wrong so when go back this information can be seen<br><br>Check illustrations with further reviews<br>Need to change candy to sweet and change it to say sweets are carbs | visual<br><br>comprehension<br><br>comprehension |
| **Protein/ fats** | Protein containing foods do not directly affect blood glucose<br>Dietary fats do not directly affect blood glucose | These need to be bold to make them stand out- this will summarise the key points without having to read whole paragraph | visual |
| **Carbohydrates** | greatest effect on blood glucose | Make bold | visual |
| **HOW SUGARS AND STARCHES EFFECT BLOOD GLUCOSE** | Graph- it is not clear that you can click on the graph and make it bigger. Knew what graph meant/ represented | Need to say click to enlarge | comprehension |
| **Digestion of carbohydrates** | Diagram is a good clear explanation, understood what it meant | | |
| **Types of carbohydrates illustrations** |  Not clear these are potatoes | Review images with other participants. Change if necessary<br>Review speed of images | Comprehension<br><br>technical |

**Figure 36 Example of review analysis**

The MAGIC programme was highly regarded by the review participants. Some of the participants' comments on the overall feedback of the MAGIC programme are highlighted in table 32. Very little negative criticism, other than that described in tables 33-37, was received.

**Table 32 Overall feedback on the MAGIC programme**

| Participant comments on their overall impression of the MAGIC programme (participant number) |
|---|
| *It's a useful tool all of this, would have made things a little easier if someone had said to me 'look here's a website, go to this when you get home.' If you've got any questions you're welcome to call us. But you know this is a useful resource for people, I do think. Because you know sometimes, you know, we don't want to call and hassle you over one little question. (1).* |
| *It's good to read their experiences especially because you obviously can bring up things that they might bring up, they might be worried about (2)* |
| *I like the way it is broken down into different section. It feels like I could, on one bus journey read one section, on the next I'll read he next. So it's nice that it's not just one long document. It's really friendly (4)* |
| *I think it's a really good website because its got, it's very comprehensive but it doesn't feel like there's too much there. Erm, the module doesn't feel like it's got too much in it, it's well structured as well and erm I personally learnt a lot. (5)* |
| *The videos are good, you haven't just got people in white coats, not that there is anything wrong with them. You've got real people in there, that's good as well. (7)* |
| *I wasn't this educated at the beginning and I think this would educate me quicker at the beginning, being diagnosed. (8)* |
| *Oh it's brilliant, I wish I had it sooner, years ago. (9).* |
| *Um, well it was easy to, easy on the eye, clear. Not just loads and loads of text that you have to read. Also diagrams and little challenges does probably get you thinking more, and you don't switch off. (10)* |

**Table 33 Omissions identified within the MAGIC programme**

| Module | Example of omissions | Example participant quotes (participant number) |
|---|---|---|
| **CFD beginner** | • No explanation that the personal experience quotes are from real people<br>• Word missing<br>• Heading from hypo flowchart missing | *It might be worth having a heading there. (1)* |
| **CFD improver** | • Missing capital letter<br>• Do not say what FIT is<br>• Time missing on mode of insulin action charts<br>• Use terms long-acting and background instead of just the agreed term long-acting<br>• Test your learning quiz does not say only tick one answer<br>• Personal experience quotes under carbohydrate section are missing a heading | *And what's the one across the bottom? Time? (5)*<br><br>*Can you click more than one? (4)* |
| **CFD advancer** | • No warning to clarify that not all the information is medical advice e.g. personal experience quotes/ videos<br>• Word missing | *You could say not all information given in the videos are, not necessarily err medical advice that we would give to our patients. I'm thinking particularly about the third guy. (7)* |
| **CFD wizard** | • No information on people with CF needing more salt<br>• Letter missing from your, says you<br>• No instructions for insulin dose adjustment chart<br>• Challenging foods video not clarified that for people with type 1 diabetes, as all the other videos are | |

**Table 34 Errors identified within the MAGIC programme**

| Module | Example of errors | Example participant quotes (participant number) |
|---|---|---|
| **CFD beginner** | | |
| **CFD improver** | • Reference to a carbohydrate table which no longer exists<br>• Wrong classification of milk as fast acting<br>• Reference to how insulin is absorbed from the arms, however the arms are not included in the illustration<br>• Comma missing from text<br>• Of should be if<br>• Use should be used<br>• Cause instead of causes<br>• Sweets are carbs- drop and drag identifies incorrectly | |
| **CFD advancer** | • Cooked and uncooked rice are labelled as the same weight<br>• Table and example for calculating carb content of weighing foods do not say the same value for potatoes<br>• Food and carb content not lined up correctly in food diary<br>• Example of calculating insulin to carb ratio- values do not agree between table and text<br>• Use of this instead of it<br>• Use instead of used<br>• Feedback Q2 and Q3 on calculating meal do not match with correct answer | |

| | | |
|---|---|---|
| | • Q6 of test your knowledge -discrepancy between text and correct answer | *What, err right. Every 30 minutes, well I'm not surprised I got that one wrong. I thought it said 2 hours earlier when I read that. I thought it said every 2 hours? (6)* |
| **CFD wizard** | • Calculation in recipe examples are wrong- lasagne and scones mixed up<br>• Dairy and alternatives and beans, pulses, fish, eggs, meat and other proteins- title and images are around the wrong way | *Make sure you've got the labels right, cause I was confused. (9)* |

**Table 35 Visual issues identified within the MAGIC programme**

| Module | Example of visual issues | Example participants quotes (participant number) |
|---|---|---|
| **CFD beginner** | • Why we need insulin illustration – words too far away from illustration<br>• Effect of food on insulin- images hazy<br>• Text quite small in some sections<br>• Use of bullet points helpful<br>• Highlight key words in bold to make clearer<br>• Not clear graph enlarged when clicked on it | *Think it could be mildly confusing in some way, unless you can combine them and just have one group. I would be wondering why is this one here? (1)*<br><br>*The writing is bolder, so that is better (2)* |
| **CFD improver** | • Did not notice well done- consider colour scheme used<br>• Highlight key words e.g. do not<br>• Not clear graph enlarged when clicked on it | *I didn't pay attention to that to be honest. (3)* |

| | | |
|---|---|---|
| | • They all contain the same amount of carbohydrate box not clear<br>• In drop or drag quiz -need a reminder if answers were correct or not for when move up and down the page. | *I wouldn't be able to remember which ones should be here and which ones should be there. So, if you had a tick or a cross over each one. (4)* |
| **CFD advancer** | • Size of print on bread label a bit small<br>• Box with answers on which food contains the most amount of carbohydrate quiz is not clear<br>• To look visually the same should have a box under each food label to summarise amount of carbohydrate<br>• Would like to see answer after each meal on carbohydrate counting activity<br>• Didn't like use of different colour to highlight section in another module | *I mean that's small though, isn't it? [..] I think that's too small to read. (7)*<br><br>*I think keeping it consistent. The way my mind works I'm looking for the same thing on each page and actually on this one you have to scroll down to see that text. And on this particular screen I hadn't scrolled down so hadn't seen it. (8)* |
| **CFD wizard** | • Table of blood glucose confusing, shading would help to make the numbers stand out | *I don't know if doing something with the graph, putting a bit of colour, or just that bit, the time bit, cause its just numbers. And for somebody who really doesn't struggle with numbers, for me it's a little bit, I'm struggling a bit. It's not clear. (9)* |

**Table 36 Comprehension difficulties identified within the MAGIC programme**

| Module | Example of comprehension difficulties | Example participants quotes (participant number) |
|---|---|---|
| **CFD beginner** | • The term respiratory exacerbation was not thought to be understandable by all- chest infections considered a more appropriate term<br>• Expand and collapse not clear throughout- click to enlarge considered clearer<br>• Images of carbohydrates- bread unclear<br>• Need to clarify that should not use hand cream prior to checking blood glucose<br>• Use of language consider changing some people to people to reflect a more common problem<br>• Didn't know what a PEG was | *It says more frequent respiratory exacerbations; do you know what it means by that?*<br>*No, what does that mean? (2)*<br><br>*I wouldn't know to click on that, I'll be trying to squint and read it. (1)* |
| **CFD improver** | • Use term sweet not candy<br>• Drop and drag- some images not clear (oil, potato)<br>• Types of carbs- images not clear (potatoes, yoghurt, rice, oats, pancakes)<br>• Quick vs slow- some images not clear (breakfast cereal, pasta, fizzy drink)<br>• DUK video- not clear what this link is, need an introductory sentence<br>• FIT website- link not clear, need to say further information can be found: | *I don't know what the hell that is. (3)*<br><br>*I don't know what that is, perhaps a potato? (*4) |

| | | |
|---|---|---|
| | • Use of language- if you do eat, reads better if you eat<br>• HbA1c- is the section about NICE targets for type 1 diabetes necessary, a bit confusing as wan to know own targets<br>• The section on the concentration of glucose in the airways was not clear and easy to understand | |
| **CFD advancer** | • Which foods contain the most amount of carbohydrates- not clear need to drag the word to the box or what some of the pictures contained<br>• How to count – not clear should use the per 100g figure<br>• Consider use of cross referencing to the exercise section for link with rounding up/down. Ask questions about if people will know the effects of exercise<br>• Carbohydrate counting activity not clear what have to do<br>• When reading the underweight section needed reminding this was for underweight and not everybody<br>• Q9 test your knowledge- the option more activity than usual during the previous evening was confusing<br>• Living with CFD one participant was having problems with the terms try and identify. Which they felt could be a more personal reflection of their experience at that time. The other two participants did not struggle with this | *I also thought that was just a roast chicken rather than a whole roast dinner so, because you can't see any potatoes in there. So I wouldn't have calculated potatoes in my head. (8)* |

| | | |
|---|---|---|
| | • Term 'get help' – 1 participant again was worried about use of language, prefer term 'receive'<br>• Test your knowledge quiz- not clear can only tick one answer<br>• To remind again what fast -acting carbohydrates are would be helpful to have some examples in the driving section | *Or is it just the wording? Try and identify which parts. I'm wondering if that can be a bit softer- I don't know. (7)* |
| **CFD wizard** | • Fish- not understanding why it says to have two portions fish per week here, what if didn't eat fish. Felt it was a bit confusing and could be removed as there was a link to the Eatwell guide.<br>• Wording in the UK Chief Medical Officer guidelines is not clear<br>• Wording why alcohol causes hypoglycaemia not clear<br>• Did not understand why a blank insulin dose adjustment diagram was in the text<br>• One participant questioned if people would know how to increase their salt intake- both participants knew how to<br>• Steroids- the picture used looked for like pancreatic enzymes | *So if you don't eat fish what would your alternative be? (9)*<br><br>*Erm I don't think so cause people would know that wouldn't they? It's quite straight forward. (10)* |

**Table 37 Technical issues identified within the MAGIC programme**

| Module | Example of technical issues | Example participant quotes (participant number) |
|---|---|---|
| **CFD beginner** | • Quality of sound in what is CFD video; one participant felt that the voice over was a bit drab and the video was formal.<br>• Personal experience videos – the voice in one of the videos is not in synch<br>• Can't see all the side headings on the screen | *This scrolly thing, I assume if you go on it, it should go up and down, but it doesn't. (1)* |
| **CFD improver** | • Types of carbohydrates- illustrations move too quickly<br>• How to use an insulin pen video- the sound quality goes up and down<br>• Relationship between CF & CFD – the videos in this section could not be opened in the programme, videos loop together<br>• Personal experience quotes in hypo section move too fast to read<br>• Personal experience videos in hypo section loop together<br>• Suitability of programme for tablets/ phone highlighted | *They move a bit fast. (4)* |
| **CFD advancer** | • Personal experience videos in carb counting section- the sound of the second video is low | |

| | | |
|---|---|---|
| | • Music on introduction to carbohydrate counting video made by Norfolk and Norwich was a bit annoying for one participant<br>• One mention of considering the use of subtitles on videos<br>• Quotes in living with CFD section move too fast<br>• Format of side panes is grey, could there be a way of matching it to module colour | *Didn't quite get the end of this. Doesn't want me to read that does it? (6)* |
| **CFD wizard** | • Managing insulin in daily life- personal experience quote move too fast to read | *Slightly distracted, maybe if it (quotes) was underneath? (10)* |

One other comment, received from two participants, was to consider the use of a glossary for key terms and definitions.

## 5.5 Discussion

The findings from this study are discussed in relation to the research question "what does a web-based education programme for people with CFD need to contain?" Clinical implications of the research findings, methodological issues and ideas for further research will then follow.

The precise content for DSME programmes is not specified in NICE guidelines (NICE 2016). The content for the MAGIC programme was guided by four key areas of self-management: insulin, managing nutrition, blood glucose and 'what is CFD?' which were identified from stages 1a and 1b, and the stakeholder development group. DSME programmes, that meet NICE criteria (NICE 2016), including DAFNE (DAFNE Study Group 2002) and BERTIE (Everett et al. 2003) also focus on insulin, nutrition and blood glucose as key areas, however as they are aimed at people with type 1 diabetes they have a greater emphasis on healthier eating principles, exercise and diabetic complications including diabetic ketoacidosis (DKA). They both also explore pregnancy and contraception which has not been addressed in the MAGIC programme. BERTIE (2019) has an extensive 'streetwise' section that covers issues not discussed in the MAGIC programme such as: drugs, school, leaving home, tattoos and piercing. DAFNE (DAFNE Study Group 2002) is delivered in a group setting whereas BERTIE (Everett et al. 2003, BERTIE 2019) is the only available type 1 DSME programme available to groups and online. There are currently no evidence-based DSME programmes available for people with CFD. The MAGIC programme is evidence-based and supports the development of the core self-management skills required to successfully manage CFD via a web-based learning platform.

In agreement with the MRC Framework (Craig et al. 2008) and to meet NICE criteria (NICE 2016) the MAGIC programme was developed with sound theoretical underpinnings which included self-efficacy theory, self-management framework for

chronic illness and adult learning principles. These theories are commonly used in DSME (chapter two) and were identified as being the most relevant to meet the learning needs of this population. At a later stage of research to demonstrate effectiveness of the MAGIC programme self-efficacy will need to be considered as an outcome measure.

The review of the MAGIC programme was designed to: explore understanding and ease of use, identify missing information, establish any visual issues and detect problems with the information or instructions provided. Ten people with CF took part in these reviews; six of these had CFD. Overall the programme was well received by the participants. The reviewers identified omissions, errors, technical issues, visual problems and comprehension difficulties within the programme. All errors and omissions were discussed with the digital content manager and the necessary amendments were made. All visual issues were discussed with the digital content manager and all except the format of the carbohydrate counting meal activity were changed. One participant did not like the answers to the carbohydrate counting meal activity appearing at the end of the activity, they would have liked to see the answer appear after each meal instead. The programme that was used to write all the quizzes for the MAGIC programme did not have the facility to give answers after each individual question. This being the opinion of a single participant, and the overall quality of the programme not being significantly affected by this format, the decision was made to leave it as it was. Changes could be made after future reviews if deemed appropriate.

The comprehension issues identified were considered very carefully. Where difficulties with clarity of images were highlighted, these were changed. Instructions on how to enlarge charts and graphs were made clearer with use of 'click to enlarge labels.' In the alcohol and HbA1c sections, where clarity was needed because the text was complex and difficult to understand, modifications were made using the suggestions given by the participants. Where comprehension issues were identified in the living with CFD section further advice was sought from patient members of the stakeholder development group to improve the language used.

All technical issues concerning sound, the movement of images/quotes and videos were reviewed with the digital content manager. Due to inconsistencies in sound the narration for the how to use a blood glucose monitor video and how to give an insulin injection video was re-recorded. One participant felt the voice on the what is CFD video was a bit drab and considered it to be quite formal although they then acknowledged it was meant to be. The video was shown to four other participants during the reviews and none of them commented on this. It has also been shown the stakeholder development group and PhD project management group during its creation, with no further comments received. The decision was therefore to go with the consensus and to leave the video as it was. All moving images and quotes were either slowed down or stopped from moving so that they could be read more easily and did not distract the participant from the content on screen.

The review generated two comments about the use of a glossary. The idea was considered, but has not been included in the MAGIC programme at present, due to prioritisation of all the other amendments that needed to be addressed in the timeframe. However, opinions from the stakeholder development group will be sought to review the need for a glossary as a future addition to the MAGIC programme.

Using the principles of co-design, the stakeholder development group created an evidence-based DSME programme for people with CFD. This has been subject to review to identify any issues with face validity in a small group of adults with CF/ CFD. The majority of the issues identified were subsequently amended. The MAGIC programme is unique as no other programmes for people with CFD of its kind exist. It is now at the stage that further research is needed to evaluate its impact on the key CFD behaviours of managing blood glucose, insulin and nutrition.

### 5.5.1 Strength and limitations

The MAGIC programme is grounded in evidence generated from a qualitative systematic review, qualitative interviews and shared experiences of people with CFD and healthcare professionals expert in the management of CFD. The content of the programme is therefore relevant to the users it was designed for. Deficits in CFD health

service provision was identified in chapter three and four, the MAGIC programme could be integrated into service improvements to provide educational and self-management support to people with CFD.

The MAGIC programme was developed through the process of co-design. The main benefits of using a co-design design approach are: the end product reflects customer needs, it generates useful information and it creates a feeling of involvement and ownership (Bradwell & Marr 2008). With co-design there is equality in the contributions from all involved (Bradwell & Marr 2008). Maintaining equality was an additional challenge during the use of videoconferencing for the stakeholder meetings. The appointment of a 'link person' to monitor links with people at home ensured equality. Alternatives methodologies to co-design such as participatory action research and experience-based co-design were considered but not deemed appropriate as they do not focus on intervention development.

The MAGIC programme has been designed to meet NICE criteria (NICE 2016) however delivery by trained facilitators, quality assurance and audit of outcomes will need to be addressed in future research. As recommended the development of the MAGIC programme was driven by relevant theory appropriate to the needs of people with CFD (NICE 2016). Self-efficacy is a particularly important factor that has been identified in influencing diabetes self-management behaviours (Mishali et al. 2011) and was very influential on the MAGIC programmes staged approach to learning.

The participants who reviewed the MAGIC programme were known to me in my previous role as a healthcare professional. This may have influenced the findings and also challenged the researcher-clinician role (Hay-Smith et al. 2016). Before starting the interviews, I explained to the participants that the purpose of the interviews were to review the MAGIC programme, and I was there as a 'researcher' and would not comment on their knowledge or self-management practices. Participants are often said to have difficulty in differentiating between a clinical consultation and research contact (Hay-Smith et al. 2016). Despite discussing my research role some participants still sought opinions as to if what they were doing was correct, particularly if for

example their method of carrying out a task was different to the method the programme used. For example, one participant calculated carbohydrates in a different method, this was due to their more advanced level of understanding and maths skills. In another interview a participant attempted to seek advice for their parent with diabetes. I remained non-judgemental throughout the interviews and where possible avoided clinical discussions. A clinician-researcher cannot completely discard their clinical role, what is important is the acknowledgement of this role and exploration of challenges created by it (Haye-Smit 2016).

Another limitation of being known to the research participants in a clinical role is social desirability bias. This is the tendency for participants to respond to questions in a way that is thought of more favourable so they present themselves in a more positive way (Neeley & Cronley 2004). The participants may therefore have been less critical and therefore more likely to only say positive things. I don't believe this happened, to a great extent, as participants gave both criticisms such as the images on the effects on food on insulin were hazy, voice over on one of the vides was drab, text is quite small in some sections along with positive comments such as the use of bullet points is helpful, the videos are good and 'oh it's brilliant.' Many important issues were identified within the participants reviews which helped inform necessary modifications to improve the MAGIC programme.

This sample size for the reviews was small compared to some studies using cognitive interviewing techniques, where sample size ranged from 19-40 participants (Banna et al. 2015, Fernandes Davies et al. 2016, Park et al. 2017, Anderson et al. 2018). One study, a pre-pilot usability testing of a web-based programme conducted cognitive interviews with 4 participants (Berry et al. 2010). The sample size of this study, to review the face validity of the MAGIC programme, was deemed relevant to the study design, allowed for two or three participants to review each module and was feasible in the timeframe allocated for this stage of the study. The cognitive interviews in this study were fairly lengthy (median 107 minutes). In studies that reviewed surveys or questionnaires cognitive interviews were shorter, lasting 40-60 minutes (Carbone et al. 2002, Fernandes Davies et al. 2016, Park et al. 2017). In one study that used cognitive

interviewing to evaluate a web-based decision support system, for men with prostate cancer, it took 60-90 minutes to complete the programme and cognitive interview (Berry et al. 2010). Therefore, more complex interventions/ resources such as web-based programmes, which are often multidimensional, take longer to review. The sample did not contain any young people with CFD, three of the 21 letters of invitation to participate were sent to people less than 26 years old but they did not respond. One of the patient members of the stakeholder development group was younger than 26 years, so there was some representation and experiences within the MAGIC programme development within this age range. Having younger members of the group may have had different influences upon the programme and its content. Particularly adolescents as they have specific needs associated with seeking independence such as going to university, leaving home or starting work which have not currently been addressed.

CFD is a complex disease to manage, the MAGIC programme was informed by four key components which were identified as being significant in the self-management of it. This meant that not all areas of CFD management were included within the MAGIC programme. Psychosocial and emotional support have not been addressed in detail, but their importance in CFD self-management has been acknowledged. The stakeholder development group recognised the importance of psychosocial and emotional support for people with CFD and therefore incorporated as much guidance, support and personal reflections where it could. The MAGIC programme contains two sections on living with CFD which includes personal experiences quotes/videos and identifies mechanisms for further support. Usability testing of a web-based programmed for adolescents who had a solid organ transplant, demonstrated patient experience videos were well received due to their trustworthiness and reflectiveness of real patient's experiences although their impact on self-management was not reviewed (Korus et al. 2015). The use of personal experiences throughout the MAGIC programmes reflects what it is really like to live with and manage CFD and these messages will provide some support by learning through shared experiences. Online support groups have been shown to help young people with CF share their feelings and

experiences to help support self-management (Kirk & Milnes 2016). Personal experience videos and quotes are the MAGIC's programme first step in this process. Through further usability testing other forms of communications such as chat boards or discussion groups may be considered. It is also important to consider that National guidelines recommend that all people with CF have access to a psychologist as part of their routine care (NICE 2017), so their emotional health and well-being is regularly assessed. The programme facilitator will also be a means of contact with the participants and their role in support whilst undertaking the programme will need evaluating.

Other components not included in the MAGIC programme were pregnancy, contraception, transplantation and moving away from home/starting university. This does mean there were not considered relevant they were not regarded as key factors, by the stakeholder development group, that affected the majority of people with CFD. In the future additions to the programme may be considered.

### 5.5.2 Implications for practice

The MAGIC programme is an extensive resource to facilitate the self-management of CFD. It is versatile programme designed to enable people with CFD to acquire the necessary skills and knowledge to manage their CFD. Its web-based delivery means it can be used at home or other convenient times, thus avoiding the need to travel to the hospital more than necessary. The MAGIC programme could be integrated into the delivery of CFD care as a resource for people newly diagnosed with CFD or for those with established CFD who could benefit from further education. The MAGIC programme may also be of benefit to healthcare professionals working in CF to try to improve the knowledge deficits observed among healthcare professionals about CFD (chapters three and four).

This study demonstrated the face validity of MAGIC programme however to establish its effectiveness further research is needed. A feasibility study is required to:

- Identify how the MAGIC programme is used in the 'real-world' situation. This includes: delivery of the programme, the use of the personal log function, food and

blood glucose diaries and secure log ins which were not reviewed within the scope of this study

- Determine which outcome measures are appropriate and subject to change
- Establish the role of the facilitator in delivering the MAGIC programme
- Explore recruitment and retention rates

## 5.6 Conclusion

Through the process of co-design this study has developed a unique evidence-based self-management education programme for people with CFD. The MAGIC programme was highly regarded by people with CFD, involved in this study. Its design as a web-based programme allows for more timely and appropriate delivery of self-management education. Further studies are however required, to demonstrate effectiveness, before the MAGIC programme can be used in routine clinical practice.

## 5.7 Research Outputs

The development of a self-management education programme for people with CFD. Oral presentation at adult and paediatric CF research away day, Royal Brompton & Harefield NHS Trust- (12th July 2019).

Collins, S., Jones, A., Woodward, S. & Sturt, J. (2019) P403 The experience of co-designing the MAGIC programme for people with CFD. *Journal of Cystic Fibrosis***18**, S171. Poster presentation at European CF Conference, 5-8th June 2019

PhD project management group meeting 3, 4th January 2019- presentation of the development and review of the MAGIC programme.

# 6 Discussion

This study has developed an evidence-based self-management education programme for people with CFD. This chapter summarises key findings and discusses these in relation to the research questions, relevant literature and theory. This is followed by a review of the strengths and limitations of the research conducted, the implications for clinical practice and recommendations for further research. Figure 37 summaries how the MAGIC programme development was informed by the research questions and the key themes identified in stage one.

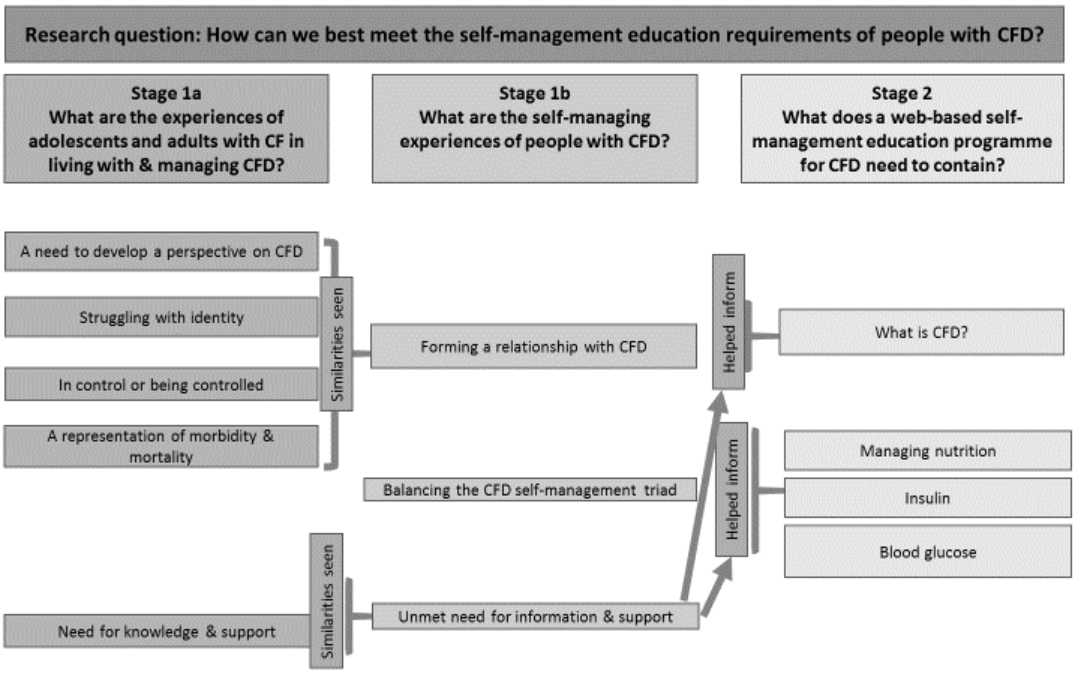


**Figure 37 Key themes and how they informed the MAGIC programme development**

## 6.1 Review of key findings

This study has demonstrated five keys findings: the response and adaptation to CFD included learning about CFD and its management and developing support and relationships; trying to balance the relationship between CF and CFD involves many complexities; there are deficits in service provision and support for people with CFD; the MAGIC programme focussed upon the physical needs of people with CFD; and

lastly the MAGIC programme was developed as a staged approach to learning. These will be discussed in more detail.

### 6.1.1 The response and adaptation to CFD

The response to chronic illness has been described as first responding to effects of the illness followed by the processes of adaptation and management (Charmaz 1995, Paterson et al. 1998). Adaptation to, and management of, chronic illness are simultaneous and interdependent; they are the processes occurring as the individual tries to come to terms with the long-term impact of their illness and include learning about the illness and its management, acquiring support and establishing relationships (Paterson et al. 1998). The response to CFD was no different to that of other chronic illnesses and these processes of adaptation and management were observed in this study. Initially the diagnosis of CFD was associated with many negative emotions such as confusion, shock and fear which changed as people began to adjust to life with CFD. As people began to learn to incorporate CFD into their lives issues which challenged identity, coping mechanism, understanding, control and the constancy of CFD were seen. These findings are not unique to adaptation to CFD as they have also been seen in people with type 1 diabetes (Schur et al. 1999, Celik et al. 2015, Robinson 2015, King et al. 2017). For CFD the focus of management, for participants, was about the essential components required for the day-to-day management of CFD; these included self-management skills, knowledge and balancing CF and CFD. Throughout the clinical course of CF, due to changes in health, management and adaption are simultaneously evolving.

When exploring the experiences of people with CF it is important to consider that these individuals have experienced healthcare systems from a very early age. They have attended hospitals and clinics on a regular on-going basis for medical treatment and care and they are experienced at managing complex daily treatment regimens. The development of diabetes, for them, is a second chronic illness. Experiences developed while living with one chronic condition and familiarity with healthcare systems can help the management of subsequent illness (White et al. 2016). This was observed in those people with CFD who incorporated CFD management into their CF

routines and thus experienced less management difficulties. For those who managed the conditions separately there was more disrupted routine and less ability to cope.

CFD had a variable impact on individual day-to-day life and this was influenced by changes in health status, ability to cope and level of understanding. The 'Shifting Perspective Model of Chronic Illness', has been described (Paterson 2001). This model views living with a chronic illness as an ongoing and continually shifting perspective in which either wellness or illness is in the foreground. Wellness in the foreground perspective suggests the management of the chronic illness itself is paramount in order to keep ill health away. Paterson (2001, p 24) states "the illness requires attention in order not to have to pay attention to it." This perspective is not static and reflects an individual's needs and situation. This perspective is applicable to the management of CF which requires extensive daily treatment regimens to maintain health where the focus is on wellness rather than illness. However, the development of CFD further increases this burden of treatment and monitoring. CF was perceived as having more impact on physical health than CFD and therefore affected the ability to complete daily activities, which would be related to its impact on the respiratory system and the associated effects of reduced lung function.

### 6.1.2 Trying to balance the relationship between CF and CFD

Living with CF and CFD was regarded as a complex-multi-dimensional process where the two conditions were intertwined. The perception of CFD, was it seen and managed as part of CF or as a separate entity, directly influenced management. This study proposed an integrated model of management based around the presumption CFD was regarded as part of CF. When viewed as one condition CFD will be managed as part of routine CF care; all treatments, therapies and self-care strategies such as behaviours, beliefs and coping mechanisms, are managed together; what affects CFD will directly impact CF and vice versa. In middle and older aged adults with diabetes the presence of a discordant conditions such as cancer, arthritis or lung disease were associated with lower diabetes prioritisation and lower self-management ability (Kerr et al. 2007). Although the age of this diabetes population is different from a typical CFD population, it demonstrates conditions that are not considered related to each

other are managed separately and this influence self-management. CFD and CF should be managed together, however further work is needed to establish the best way to manage gaps in service provision to support this.

People with CFD view their CF as the most important illness because of the recognition they will die from CF. They therefore prioritised the management of their CF. Prioritisation of disease management is not uncommon in people with multi-morbid conditions. A study of heart failure self-management practices in people with multimorbid conditions demonstrated that individuals also had to prioritise one condition over another (Dickson et al. 2011). They based decisions upon their experiences with the different conditions, for example it they had experienced a life-threatening episode due to complications of heart failure they would prioritise this. Self-management decisions were informed by both experiential beliefs (the behaviour is easy/difficult) and evaluative beliefs (the behaviour is important/unimportant) (Dickson et al. 2011). People with CF found the effects of not doing CF treatment were more noticeable and thus more important to them than those of CFD. In adults with diabetes, the severity of co-morbidities influenced an individual's ability to focus on diabetes and to perform self-management tasks; with diabetes viewed as a lower priority and its self-management tasks considered more difficult (Kerr et al. 2007). Diabetic renal disease is a co-morbidity associated with diabetes. For many of these people, with diabetes, the development of diabetic renal disease was a sign to them that they needed to regard their diabetes more seriously, specifically dietary and treatment regimens (King et al. 2002). Generally, there was an avoidance of thinking about the future progression of their renal disease and/or diabetes as this was considered unhelpful. There was a sense that individuals carried on to the best of their ability, living from day-to-day, not dwelling on their own mortality. Many were more concerned with the development or exacerbation of other complications, particularly blindness. Participants with CFD, in this study, were aware that they were at risk of diabetes related complications, especially eye damage, but they didn't ruminate on these, they just put their thoughts concerning this in the background. People with renal disease and diabetes will have frequent contact with other people with the same

condition through hospital appointments and dialysis sessions. They may have therefore had more exposure to people with significant diabetes related complications such as blindness. People with CF are advised against face-to-face contact with other CF patients and are therefore less likely to see people with CFD and diabetic related complications. They are therefore likely to worry less about the risk of blindness and other complications.

A qualitative study, of adults in Taiwan, explored experiences from the diagnosis of diabetes through to haemodialysis (Yu & Tsai 2013). Diabetes to most participants was identified as a disease that was chronic and long-term, and upon its onset it was not felt to be an immediate risk of death. The perception of diabetes symptoms affected attitude towards disease control; with self-management neglected unless symptoms were apparent. People with CFD were also not concerned with the risk of death from diabetes. They however didn't appear to neglect their CFD, it was more an issue with balance and prioritising what was important to them at that particular moment in time. With CF often being prioritised because the effects of not doing CF treatments were more noticeable and immediate. Diabetes needs to be considered in context, in these Taiwanese adult's cultural beliefs informed decisions relating to changes to diet and lifestyle, with changes not being made as symptoms were not evident.

Since the late 1990s the term cystic fibrosis related diabetes has been used to describe diabetes in people with CF. Prior to this it was referred to as diabetes of cystic fibrosis or cystic fibrosis diabetes mellitus. In view of the recognition that CFD is caused by CF and to strengthen the relationship between these two conditions a name change from cystic fibrosis related diabetes to cystic fibrosis diabetes was proposed. In 2014, international consensus guidelines recommended that diabetes diagnosed in post-transplant patients changed its name from new onset diabetes after transplant (NODAT) back to post transplant diabetes mellitus (Sharif et al. 2014). This was as a recognition that not all cases of diabetes diagnosed post-transplant are new; the diagnosis may have been missed pre-transplant due to ineffective screening programmes. Therefore, the name post-transplant diabetes mellitus avoids any misconceptions and describes diabetes diagnosed in all post-transplant patients.

Diabetes has historically gone through many name changes including juvenile or adult onset diabetes, insulin dependent diabetes or non-insulin dependent diabetes and the current terminology, of type 1 and type 2 diabetes. The diabetes community are not happy with current terminology and have petitioned for changing the name to reflect the nature of diabetes (Hoskins 2018). Diabetes associated stigma is another driving force for changing the names used (Browne et al. 2014). In people with type 1 diabetes stigma has been seen as type 1 diabetes specific e.g. discrimination, exclusion and also stigma-by-association with type 2 diabetes e.g. blame, stereotyping (Browne et al. 2014). Similarities have been seen in managing schizophrenia, where the impact of its name is seen to be stigmatising and harmful (Lasalvia et al. 2015). Japan was the first country to change its name for schizophrenia from 'split mind' to 'integration dysregulation syndrome' (George & Klijn 2013). This resulted in better acceptance of diagnosis and a reduction in stigma. This has however not been internationally adopted (Lasalvia et al. 2015). Although people with CFD did have concerns around stigma experience particularly around giving insulin injections in public, strengthening the causal relationship and establishing the therapeutic and treatment links between CF and CFD is more important and was the driving force behind name change.

One of the challenges seen in the management of CFD was associated with the unpredictable nature of both CF and CFD. There are lots of uncertainties living with an unpredictable long-term condition like CF where the experience has been described as being like a 'rollercoaster' of fluctuations in health (Knudsen et al. 2018). This can result in day-to-day variability in clinical symptoms that in turn impact upon blood glucose levels and the management of CFD. The management of CFD is complex due to multiple factors, pertinent to CF, that affect glucose metabolism. These include: malabsorption, chronic/acute respiratory infections, glucagon deficiency, abnormal intestinal transit time, liver dysfunction and increased energy expenditure (Dyce et al. 2015). People with CFD had to continually make treatment and management decisions which changed frequently; this added to the overall complexity in CFD management.

Taking corticosteroids as a treatment of CF further added to the complexity of CFD management. This was in terms of the effects of the corticosteroids on physical

appearance and blood glucose levels and for some the psychological effects such as mood swings. There was a sense of disbelief that a treatment which was meant to be benefiting their CF has such negative effects upon them. One participant had an extremely negative relationship with corticosteroids as he blamed them for causing his CFD; this led to him boxing off diabetes and not fully accepting it. These findings are unique to the IPA study as no qualitative work looking at experiences of taking corticosteroids in people with CF exists. The impact of corticosteroids on blood glucose levels has been reported in CF; approximately one-third of adults and children with CFD found controlling their blood glucose challenging whilst taking corticosteroids (Millington et al. 2014). Limited qualitative research exploring the experience of taking corticosteroids in other illnesses also exists. Haematology patients have reported physical symptoms of corticosteroid use to include: weight gain, extreme hunger, swollen face, fluid retention, visual disturbances, bruising and flaky skin. They described these physical symptoms as distressing and challenging and they exacerbated the psychological difficulties experienced with corticosteroid use (McGrath & Holewa 2010).

### 6.1.3 Deficits in service provision and support for people with CFD

Healthcare professionals were the first point of contact for the participants, however there was a requirement for them to be knowledgeable about CFD. There are currently only 2026 adults in the UK on treatment for CFD (Cystic Fibrosis Trust 2018a), CFD is therefore a rare condition and many healthcare professionals would have had limited exposure to people with CFD throughout their training and work.

A general lack of support for people with CFD was observed. People with CFD wanted to feel they had been listened to and there was some awareness from the healthcare professional of how they managed CFD in the context of their life. In people with type 1 diabetes communication with healthcare professionals also showed a lack of insight into the recognition of the person rather than the disease (Spencer et al. 2010, Robinson 2015), with focus on blood glucose control or HbA1c rather than the individual (Robinson 2015, King et al. 2017). Poor communication from healthcare

professionals can hinder type 1 diabetes self-management (Spencer et al. 2013, Robinson 2015), impact upon attendance at appointments (Celik et al. 2015) and affect psychological well-being (Robinson 2015). Support and advice from healthcare professionals to people with all types of diabetes needs to be meaningful, consider the wider context of the disease process and be delivered in a caring and considerate manner incorporating the individual in care planning.

Healthcare professionals and people with diabetes may not share the same management goals; with clinicians focusing on achieving control in terms of blood glucose levels. Where a goal of a healthy balance may be a considered as a more appropriate representation of individuals' willingness to engage in diabetes self-management (Paterson et al. 1998). This focus on balance rather than control takes the emphasis away from issues surrounding concordance. The focus is on the individual and valuing their insight into their diabetes management; how they understand and achieve balance in their lives whilst appreciating that diabetes is a dynamic disease and control can change with life events and changes in health status.

The CFD clinic was a major drawback in the provision of CFD care within this CF centre. The major limiting factors was time. The CFD clinic was held on a different day to the CF clinic and only addressed CFD management with very limited CF respiratory associated care provided. This meant patients had to attend an additional clinic for their CFD care. Separating out the clinics this way was delivering a message that they are two separate condition whereas the findings from this study demonstrated the need to treat harmoniously. People living with multiple co-morbid conditions struggle to integrate the fragmented information they get from different clinicians into their disease management, which leads to inadequate self-management and the increased risk of poor outcomes (Dickson et al. 2011).

### 6.1.4 The MAGIC programme focuses upon the physical needs of people with CFD

Essential key components to the self-management of CFD were identified as the CFD self-management triad, namely: insulin, blood glucose and nutrition. Uncertainties

regarding insulin were problematic and reflected deficits in knowledge and understanding and were further exacerbated by the unpredictability of CF and CFD. Blood glucose monitoring was considered the most intrusive element of CFD management and hence had a significant impact upon daily life. There was variability in the experiences as to what was achievable for individuals in terms of blood glucose management. Difficulties in balancing diet and controlling blood glucose were seen, with a need to improve knowledge of the role carbohydrates in CFD and how to better manage nutrition.

Another essential component to self-management which was identified was the lack of understanding about what CFD is, what causes it and how it interacts with CF. What is key in order to improve the self-management of CFD is the need to develop a greater understanding of CFD along with the provision of realistic, practical and helpful education and support on how to balance the CFD self-management triad. Being informed with knowledge to facilitate decision making, along with the provision of reliable information and the option to participate in decision making were deemed essential to help adults manage their type 1 diabetes (Jull et al. 2016).

The stakeholder development group considered these keys findings and supporting evidence, along with their personal knowledge and experiences of managing CFD, to inform the MAGIC programme development. The CFD self-management triad addressed the physical needs of someone with CFD; these are the fundamental requirements that are needed to effectively manage CFD. Ultimately this influences survival because the major driver for treating dysglycaemia early in CF is to improve or optimise nutritional and respiratory status which influences morbidity and mortality (Bridges et al. 2018). To address the identified deficits in knowledge and self-management skills, the stakeholder development group agreed with the evidence and supported the focus of the programme around the CFD self-management triad and the understanding of what CFD is. The stakeholder development group remained mindful of the emotional needs of people with CFD throughout the MAGIC programme development and incorporated guidance, support and personal reflections through the use of personal experience videos and vignettes from people with CFD. It also

remained true to the focus of the programme on managing abnormal glucose in CF. Primarily the MAGIC programme is an educational resource designed to facilitate self-management and its key focus was to address these identified deficits, so people can develop skills to effectively self-manage their CFD.

NICE (2015) do not specify what core topics should be included as part of a DSME programme. The American Diabetes Association's National Standards for DSME identify core topics for DSME, these include: nutritional management, physical activity, diabetes disease process, medical treatments, blood glucose monitoring and how it is used to inform self-management decisions, developing personal strategies to promote health and address psychosocial concerns, and preventing, identifying and treating complications (Beck et al. 2017). They however suggest content should be adapted to individual requirements according to type of diabetes, age, culture, health literacy, numeracy and co-morbidities. Thus, the MAGIC programme was developed to address the specific needs of people with CFD.

### 6.1.5 The MAGIC programme was developed as a staged approach to learning

The MAGIC programme is a unique self-management education programme for people with CFD. No similar programmes for people with CFD exist. It has been designed around a staged-approach to learning, which develops skills, knowledge and experiences in CFD management, to improve self-efficacy. The MAGIC programme consists of four modules: CFD beginner, CFD improver, CFD advance and CFD wizard. The stakeholder development group decided the CFD beginner module would be regarded as the basic level of knowledge and skills that someone with CFD would require. The modules would then increase in complexity with the final module, CFD wizard, being regarded as most advanced level of knowledge and skills that someone with CFD would require. The stakeholder development group did not make recommendations that everyone with CFD should complete all four modules; this would be decided between the programme facilitator and the person with CFD and therefore based on individual learning requirements and ability.

In a recent systematic, review eight key components of effective chronic condition self-management education interventions for children with asthma, cystic fibrosis, and diabetes were identified. Having a structured and sequential programme was considered one of these (Saxby et al. 2019). The structured and sequential programme, as in the stage approach to learning adopted by the MAGIC programme, facilitates learning by building upon the skills and knowledge previously learnt and developed. Another systematic review identified five main issues influencing the ability to self-care in people with type1 and type 2 diabetes: communication, education, personal factors, support and provider issues (Wilkinson et al. 2014). They also identified sequential learning as key part of education. For knowledge to be sustained the scaffolding of education was deemed important, this is learning by building upon previous knowledge or education, incrementally. Structured and sequential learning are thus identified as key to self-management.

## 6.2 Strengths and limitations

CF and CFD research are very medically focussed with limited qualitative research within this field as to how these co-morbidities are experienced. However, the experiences seen in managing other co-morbid conditions has demonstrated some similarities with the experiences of managing CF and CFD (King et al. 2002, Kerr et al. 2007, Dickson et al. 2011, Yu & Tsai 2013). This included: prioritisation, where one disease was considered more important than the other (Kerr et al. 2007); the severity of the illnesses informing treatment and self-management, with priority given to the most severe illness (Kerr et al. 2007, Yu & Tsai 2013) and the influence of previous experiences and personal values on self-management decisions (Dickson et al. 2011). People with other multi-morbid conditions have also been shown not to be concerned with the risk of death from diabetes (Yu & Tsai 2013) or think about future progression of it (King et al. 2002).

Although the experiences of living with CF and CFD share similarities with other multi-morbid conditions there is a deficit in CF research concerning the emotional experiences of people with CFD. The focus of future work should aim to develop a

greater understanding of emotions to help inform the development of appropriate psychosocial support and resources for people with CFD.

Interpretation is a key feature of meta-ethnography and IPA. A criticism received from journal reviewers for both the meta-ethnography and IPA study concerned not going beyond description in some of the data analysis. Data analysis, for both the meta-ethnography and empirical study, were re-worked on many occasions to, where possible, achieve a greater sense of meaning. More descriptive methods of analysis could have been used instead such as qualitative thematic synthesis or thematic analysis but the aim was to go beyond purely describing and summarising findings. It was important to demonstrate meaning in experiences, particularly with regards to context of trying to live with and manage both CF and CFD; this would have been lost with more descriptive methods.

IPA recognises the role of the researcher in making sense of participants' experiences (Shinebourne 2011). Heidegger's work acknowledged that researchers' fore-conceptions (assumptions and ideas) contribute to the interpretative process but suggests priority should be given to new things instead of the researchers' preconceptions (Smith et al. 2009). Thus, throughout the research process there was a need to remain mindful to any assumptions emerging while trying to engage with the new. I have been working in CFD for a significant amount of time and had an establish professional relationship with some of the participants. I had to put my thoughts, feelings and knowledge about them aside and engage with them, at that moment, in that context. A limitation of this was that I knew some of the patients well, and had my own understanding of them, their life and disease management. I approached the interviews trying to put these assumptions aside and sought clarity of their experiences by asking prompts during the interviews rather than being guided by my assumptions or prior knowledge. When participants made comments about colleagues or the service I knew I had to depersonalise these and not use my knowledge or experience to inform my interpretations. This process of challenging my pre-conceptions continued throughout the study.

Challenging personal interpretations are vital to ensure rigor in qualitative research (Toye et al. 2014). Therefore, the use of more than one researcher in the meta-ethnography and IPA study contributed to increasing rigor. The different professional backgrounds, knowledge and experiences of the research team offered different perspectives and challenges to the interpretative process. The key findings from each stage were also discussed with the PhD project management group, which further contributed to enhancing the credibility of the findings.

Hay-Smith et al. (2016), in a systematic review of clinician-researcher dual- role experiences, advocates "once a clinician, always a clinician," suggesting the clinician-researcher cannot completely discard their clinical role and the dual role created requires acknowledgement (Hay-Smith et al. 2016). I was known to the study participants as a CF specialist dietitian and for some this relationship had existed for a significant number of years. Indeed, one of the reasons I became a researcher was through my clinical experience. Consciously and subconsciously the association between the researcher and participant reflects that of the clinician-patient relationship (Hay-Smith et al. 2016). I always remained mindful to the possibility of the existence of dual roles within the generation of research data in this study. Dual roles of clinicians and researcher can pose many challenges throughout research; these are classified into two major themes: 1) research role can involve patterns of behaviour typical of a clinical role and 2) developing connections that start to resemble a clinician-patient relationship (Hay-Smith et al. 2016). Hay-Smiths' framework is very comprehensive and provides guidance for when struggling with the ethical and methodological challenges of dual roles. Pertinent to my research was the sub-theme from theme one that concerned clinical queries. The IPA interviews did not pose any challenges to my clinical relationship with the participants as none of them sought clinical advice or clarification and I was mindful to be non-judgemental of their experiences and management strategies. In the cognitive interviews, reviewing the MAGIC programme, more participants sought clinical opinions from me. This tended to be reassurance if what they were doing was correct. For example, one participant used a different method to calculate their carbohydrate portions, which they had developed

through experiential learning. They had merged the three steps taught in the MAGIC programme into one step, I made the informed decision to reassure them what they were doing was correct and explained how it related to the more 'simplified' method taught in the MAGIC programme. One participant in the MAGIC reviews asked me a clinical question that was related to other health issues they were experiencing, because they were post-transplant they were not seeing a not seeing a dietitian regularly. I therefore made the clinical decision to address their questions, once the interview was over. I had the knowledge and experience to decide if answering clinical questions that occurred during the research process were applicable and made decisions appropriately so they did not impede the research process. Hay-Smith (2016) acknowledges that some researchers are happy to answer, 'simple and reasonable' questions, however what this means depends upon personal interpretation. I would not be prepared to engage in what I interpret as more clinical management decisions such as changing insulin doses, reviewing blood glucose levels or making dietary changes during research interviews. I would, if required, arrange appropriate clinical follow up, for the participants, to address these issues. The clinician- researcher boundaries are not clear cut and I believe I have a moral obligation to ensure duty of care, this could be referring onto other clinicians or where appropriate providing assistance. What is important is the awareness that dual roles can exist but that they do not necessarily result in negative effects on the research process and outcomes (Hay-Smith et al. 2016).

Another limitation of being known to the research participants in a clinical role included social desirability bias. This is the tendency for participants to respond to questions in a way that is thought of more favourable so they present themselves in a more positive way (Neeley & Cronley 2004). This is comparable to the sub-theme in Hay-Smith's framework of suspicion and holding back; participants may be limiting what they share (Hay-Smith et al. 2016). Criticising care, people or services is a reflection of participants not-holding back. I have considered that participants may not have been fully open to me due to my dual roles of a researcher and a clinician. They could have retained a fear that their responses could impact upon their clinical care or

their experiences may be judged or criticised. Even though this was discussed as part of the consenting process prior the interviews. However, many criticisms were revealed in the study particularly concerning deficits in service provision and support but also about the face validity of the MAGIC programme. Overall a diverse variety of responses were obtained in all parts of the study, which have been used to strengthen the understanding of the experiences of managing CFD and establish face validity of the MAGIC programme.

The findings from this study are from one adult CF centre. They may not be completely transferable to other CF centres where different practices in the management of people with CFD exist. There is also the need to be mindful that the participants were self-selecting. Those who came forward for interviews in the IPA study are more likely to be having positive experiences although in the case of one participant anger and dissatisfaction appeared a motivator to participate. Those who participated in the MAGIC reviews were more diverse as they demonstrated variability in knowledge and skill mix. In both stages, with the exceptions of those who didn't have CFD, the participants had lived with CFD for many years. The processes of adaptation, management and self-management experiences of someone with newly diagnosed may differ from those who has had CFD for many years. A meta-synthesis of qualitative studies, exploring the diagnosis and early processes of adaptation in adults with newly diagnosed type 1 diabetes, identified five key influencing themes: disruption, constructing a personal view of diabetes, reconstructing a view of self, learning to live with diabetes, and behavioural adaptations (Due-Christensen et al. 2018). The process of adapting to the diagnosis of type 1 diabetes was significantly influenced by previous life experiences including physical, social and psychological aspects of life. These findings share many similarities to those seen in people with CFD within this study. This is particularly with regards to: the dynamic nature of diabetes necessitating constant change and adaptation, how diabetes impacts upon life is influenced by individual perception of it and the multidimensional process of learning involved in managing it. What's missing are those findings only pertinent to CFD such as the complexities in trying to achieve a sense of balance between managing CF and CFD.

Throughout this study it was difficult to recruit younger adults. This is a well-known challenge in research (Cain & McGuinness 2005, Gokee-Larose et al. 2009, Kenten et al. 2017, Peake et al. 2018). In this study, to avoid coercion, I did not actively follow up invitation to participate letters with phone calls. On reflection another member of the research team, who was not known to the patients, could have made these follow up calls to facilitate recruitment. Letters were the primary method of recruitment in this study. For future research it would be useful to consider how to target this more difficult to reach population. Consideration should have been given to adopting a more applicable method of recruitment to this age group. Social media, such as Facebook has been shown as a useful tool to recruitment in health research (Whitaker et al. 2017) and may therefore be a more applicable recruitment channel for younger adults. Convenience, incentives, accentuating benefits and advertisement have been proposed as methods to recruit young adults to studies (Corsino et al. 2013). Other suggestions to improve recruitment of young adults into research include involving them early in the research process from the study design stage (Kenten et al. 2017, Peake et al. 2018). The MAGIC study has one younger adult representative on the stakeholder development group; in hindsight and for future planning this number needs to be increased. The inclusion of younger adults may have influenced some of the topics covered in the MAGIC programme, and specific management issues relevant to adolescents and younger adults such as body image, drug use (alcohol, tobacco and other drugs), puberty and reproductive health issues may require additional attention (Withers 2012). Becoming independent is important for adolescents with CF (Withers 2012) and type 1 diabetes (Spencer et al. 2010, Castensoe-Seidenfaden et al. 2017, Strand et al. 2019) and self-management programmes will need to facilitate this. As part of seeking independence young adults may move out of home, go to university or start work, these experiences would need attention in a self-management education programme. BERTIE has a separate 'streetwise' section of their website specifically aimed at adolescents and young adults (BERTIE 2019). Consideration for additional modules to the MAGIC programme could be made, if deemed necessary, in the future.

This study had a strong patient and public representation throughout, this started in the planning of the proposal, continued through to the development of the MAGIC programme and will be incorporated into dissemination of findings. Patient and public representation was imperative to this study which has designed an education programme for people with CFD which has been heavily influence by the intended audience it was designed for.

The MAGIC programme is a self-management education programme for people with CFD. It was designed to address the skills and knowledge required to manage abnormal glucose in CF. This was informed by the evidence from stage one and expertise from the stakeholder development group. The MAGIC programme does this well in a very clear, informative and accessible way, which includes many interactive and visual components to facilitate learning. Due to the focus on the education and skills required to self-manage CFD and time constraints it was unable to address every aspect of CFD care. However, that was not the aim it set out to achieve. The MAGIC programme was not designed to address all the psychosocial component or management issues of living with CFD. It normalised that emotional challenges exist for people with CFD and made suggestions for further channels of support. There is limited research looking specifically at the emotional needs of people with CFD. Diabetes distress, the emotional distress and behavioural challenges associated with living with diabetes (Fisher et al. 2015), has been shown to affect 20-30% of people with type 1 diabetes (Sturt et al. 2015). Diabetes distress influences glycaemic control and self-management behaviours such as insulin administration/omission and blood glucose monitoring (Sturt et al. 2015). Diabetes distress has not been explored in people with CFD. People with CF experienced very similar responses and processes of adaptation to diabetes as those with type 1 or type 2 diabetes, therefore there are no reasons to believe that they will not experience some level of diabetes distress. People are generally diagnosed with CF in infancy or early childhood so for many by the time they develop CFD, in early adulthood, they will have had many years to develop their understanding of their illness experience. They will therefore be familiar with managing complex daily treatment regimens. Indeed, many adults with CF view

themselves as a ‘normal’ person with CF and, despite variable health and daily treatments, try to lead a ‘normal’ life (Higham et al. 2013). This illness experience could result in lower levels of diabetes distress than that seen in people with type 1 or type 2 diabetes. Or for some the experience of a higher burden of treatment associated with CFD may result in higher levels of diabetes distress. The MAGIC programme facilitator will help facilitate the participants’ learning. They will provide support to participants, by telephone or video-conferencing, at individually agreed time points throughout the programme. This will include exploring, with the participants, any challenges or difficulties they are experiencing with the MAGIC programme or any emotional challenges they have encountered whist participating in the programme. If any significant difficulties are encountered participants can be referred back to their clinical teams. CF teams should have psychosocial support as an integral part of their service (UK Cystic Fibrosis Trust Standards of Care Working Group 2011); this includes access to clinical psychologists and an annual general mental health and wellbeing assessment (NICE 2017).

## 6.3 Implications for clinical practice

As highlighted in chapters two and three, the meta-ethnography was the driving force to reconsider the name cystic fibrosis related diabetes. The language and terminologies used had a vital role in making sense of experiences. Removing ‘related’ strengthens the link between CF and diabetes. This is important because a deficit in understanding the relationship between CF and diabetes was seen. CFD should be regarded as part of CF; it is a complication that occurs due to CF pathophysiology. The challenge now is in conveying this message to the wider CF community, at a national and international level. The first and significant step in this was the adoption of the term CFD by the CF Trusts’ CFD guideline steering group. These guidelines will be available via the CF Trust website and can therefore be widely accessed. Any communications with the CF community, via the CF Trust, promoting this document will be using the name CFD. This will help people to start to become more familiar with this terminology. The process of promoting a name change will be a slow, gradual

process taking every opportunity available to continual promote it as widely as possible within the CF community.

CFD management needs to be comprehensive, integrated into routine care, and adaptable to consider changes in health status and life events, therefore normalising CFD management as part of routine CF care. Healthcare professionals should be conveying this message from diagnosis to facilitate understanding and to inform management strategies. Guidelines recommend all people with CF are screened for CFD from the age of 10 years (Moran et al. 2010b, NICE 2017), this is a time where people with CF are informed about the risks of developing CFD. At this point early education could be initiated to increase the understanding of the relationship between CF and CFD and the importance of effective management as part of routine care.

The provision of CFD care, particularly clinics, within this study was problematic. By holding a specialist CFD clinic which had very little overlap with routine CF care (CF clinics) the centre further contributed to separating the two conditions. This service does not support managing CF and CFD harmoniously and does not help patients develop the correct understanding about the relationship between CF and CFD. It also conveys the message that only CFD specialists can manage CFD. This study demonstrated that the knowledge of healthcare professionals about CFD was often deficient therefore the educational needs of healthcare professionals requires attention. All healthcare professionals working in CFD should be educated so that they are able to provide some level of care and support to people with CFD. This way, more people within the team will be talking about CFD and taking an interest in its management which will highlight to patients the importance of the relationship between CF and CFD and incorporate CFD management into routine CF care.

The quality improvement aspects of gaps in service for people with CF and CFD therefore need addressing to improve the delivery of support, education and care. This needs to consider: time commitments, use of technology, type of support provided and educational needs. People with CFD should have an integral part in the planning of their management; this will help inform realistic care plans that consider the wider

context beyond the illness itself. Attention should therefore be paid to developing care delivery pathways, including digital, to provide continuity of care, education and support with known healthcare professionals. This will help support the integration of people with CFD in treatment and care planning.

Many knowledge deficits essential to self-managing CFD were identified. These were concerning the self-management triad- insulin, nutrition and blood glucose and an understanding of what CFD is. The education of people with CFD therefore needs addressing. The MAGIC programme has been developed to address these deficits so individuals can develop skills and knowledge to effectively self-manage their CFD. The MAGIC programme should be integrated into the delivery of CFD care as a resource for people newly diagnosed with CFD and for those with established CFD who would benefit from further education. Due to its secure web-based format people with CFD can use it in a timely and convent manner to them, thus avoiding the need for additional hospital visits. The MAGIC programme could also be used as an educational resource for healthcare professionals.

The MAGIC programme could currently be used by people with CFD however there is the conundrum of weighing up the balance between providing patient education and providing evidence-based patient education. As a healthcare professional I have a duty of care, an element of this is being able to provide disease-specific patient education to help patients self-manage their long-term conditions. However, evidence-based healthcare requires verification that such programmes or interventions are effective. This study is currently being planned, but realistically it will be at least three years before evidence of effectiveness is available. Does the MAGIC programme sit unused for that time or should it be used? In clinical practice, time for patient education is limited and often a more reactive approach to disease management is adopted. The MAGIC programme does not necessitate hospital visits and can be used by people with CFD in a more timely and convenient manner. I believe that I should start using the MAGIC programme with patients in my CF centre. The decision for this will be made on an individualised patient basis and take into consideration clinical status, glycaemic control, current level of knowledge and understanding. A huge amount of time,

resources and money has been invested in developing this unique programme with established face validity. Using the programme will help generate some user evidence, this will include: understanding what is useful or unhelpful, identify areas where more support is needed so the role of the facilitator can be more clearly established, identify problems with the programme and establish how people use the programme- do they just dip in and out or do they work through the programme from start to finish? Attendance, either previous or planned for the future, at a DSME programme is part of NHS England's criteria for the funding of flash glucose monitoring, which people with CF and diabetes are now eligible for (NHS England 2019). Flash glucose monitoring is an innovative technology that minimise the need for finger-prick blood glucose monitoring (Abbott 2019). As no DSME programmes exist for CFD our CF centre will also need to consider how we are going to address this. Could the MAGIC programme be used? It may be, for example, that we recommend all people with CFD who start on flash glucose monitoring complete the CFD beginner module. The argument for not using the MAGIC programme is that we do not know if it will be effective and upon which outcomes it may have an effect.

## 6.4 Recommendations for future research

In the twenty years I have been working in CF the characteristic of the CF population is now more clinically diverse. Survival has improved in people with CF (Keogh et al. 2019) and people are living for longer with more severe disease (George et al. 2011). Along with improvements in health and survival comes a high burden of treatment (Sawicki et al. 2009). It now not uncommon to also treat people with CF who are overweight or obese (Panagopoulou et al. 2014, Hanna & Weiner 2015), who have different management requirements. Little research is specifically aimed at this population. It would be beneficial to identify what is the role of dietary modification in the management of overweight and obese adults with CFD, this could be through a national survey looking at practices in different CF centres. It would also be helpful to understand the personal experiences of making dietary changes from people with CFD to help inform more effective management strategies. The use of CFTR modulator therapies is also impacting disease management. To inform treatment decisions it

would be helpful to establish what are the effects of CFTR modulator therapy on glycaemia in people with CFD.

CFD is the most common co-morbidity affecting people with CF, however despite earlier screening and treatment aged-adjusted mortality remains 3.5 times greater in people with CFD compared to those without (Lewis et al. 2015). Establishing the cause of mortality in people with CFD would be valuable to inform future management strategies. There is also need for further research to inform national guidelines of when should treatment for dysglycaemia in people with CF be commenced?

This study identified a deficit in qualitative studies exploring the experiences of living with and managing CFD indicating a need for further studies exploring these experiences to be undertaken. These could be further qualitative studies, or case reports which highlight CFD self-management strategies and processes to reflect what can be learnt from those with CFD who are managing or not managing CFD well. It would be particularly beneficial to explore the experiences of those newly diagnosed with CFD and the more difficult to recruit groups, such as adolescents and young adults, to identify similarities and differences in experiences.

The MAGIC programme is a self-management education programme for people with CFD, it therefore focussed on the physical needs of people with CFD. Further research should be considered looking at the emotional needs of people with CFD to inform the development of a healthcare pathway to meet these needs.

Oral antidiabetic drugs are not widely used in the treatment of CFD, because its primary pathophysiology is insulin deficiency, and studies on their use are limited (Ballmann et al. 2014, Onady & Stolfi 2016). DPP-4 inhibitors (gliptins) may be beneficial for those with CF who have a good nutritional status, early stages of dysglycaemia and demonstrate barriers to insulin therapy (Sunsoa et al. 2017, Farrell et al. 2018). Further studies are needed to identify the effectiveness of DPP-4 inhibitors on clinical and patient reported outcomes in overweight and obese adults with CFD.

We do not know what the most appropriate clinical and patient reported outcomes are for people with CFD, and a CFD core outcome set does not currently exist. A programme of research for developing a core outcome set for clinical studies in CF has been registered with the COMET Initiative (COMET 2019). The scope of which includes respiratory, gastrointestinal, nutritional, mental health and quality of life. What is not clear is if CFD is included in this scope. The development of a core outcome data set and the development of patient reported outcome measures arising from the priorities established in the core outcome set research is an essential next step. Only once a core outcome set for CFD is established then can the MAGIC programme be assessed, against relevant outcomes, in the entire CF community.

This study demonstrated face validity of the MAGIC programme however to establish its effectiveness further research is needed. The next stage of the work is a feasibility study which will run in conjunction with the development of a CFD specific core outcome set, the logic model (figure 38) gives an overview of this.

The feasibility study is required to:

- Identify how the MAGIC programme is used in the 'real-world' situation. This includes: delivery of the programme, the use of the personal log function, food and blood glucose diaries and secure logins which were not reviewed within the scope of this study

- Determine which clinical and patient reported outcome measures are appropriate and subject to change

- Establish the role of the facilitator in delivering the MAGIC programme

- Explore recruitment and retention rates

- Develop a protocol to determine the effectiveness and cost effectiveness of the MAGIC programme in a rare disease trial

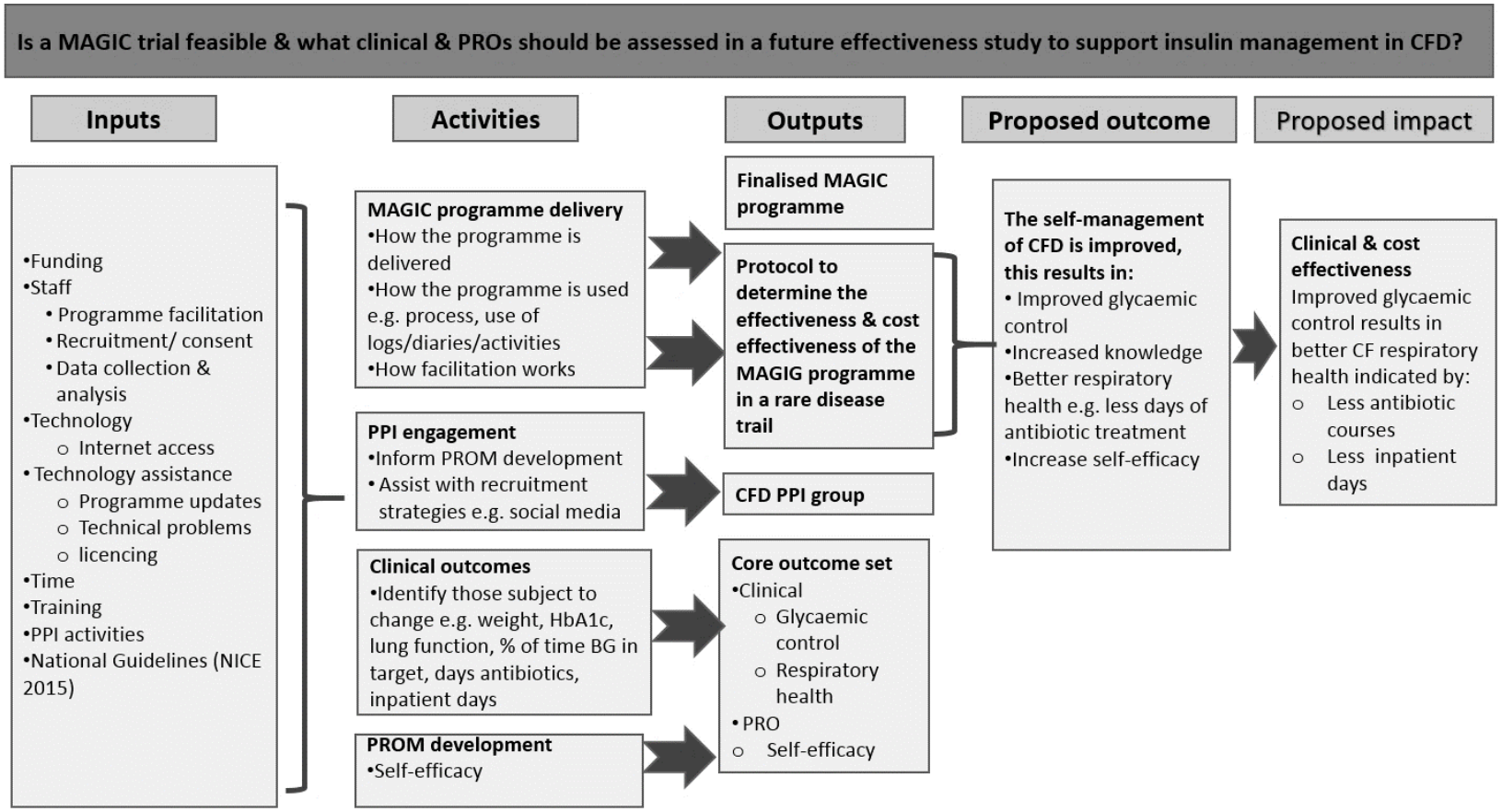


**Figure 38 Logic model**

# 7 Conclusions

This study aimed to answer the research question “how can we best meet the self-management education requirements of people with CFD?” This was achieved through two stages of work informed by the MRC Framework for developing and evaluating complex interventions.

In the first stage a meta-ethnography and an empirical IPA study provided insight into the experiences of living with, and self-managing, CFD. The response and adaptation to CFD is not completely unique and shares many similarities with that experienced by people with type 1 or type 2 diabetes and other chronic illnesses. The differences experienced related to the complexities trying to balance the relationship between CF and CFD. How CFD was perceived influenced its management. The relationship between CF and CFD is entwined, this study proposed an integrated model of management to reflect this and to promote the management of CFD as part of CF. To strengthen the causal relationship between CF and CFD, and to promote patients and healthcare professionals managing them as one condition, a name change from cystic fibrosis related diabetes to cystic fibrosis diabetes was proposed. The challenge now is to promote this within the wider CF community.

Deficits in service provision and lack of support for people with CFD were identified as limiting factors in the care of people with CFD. To provide continuity of care, education and support for people with CFD a more timely, digital and harmonious service which is integrated into routine CF care is needed.

The CFD self-management triad (blood glucose, insulin and nutrition) and a lack of knowledge regarding what is CFD were considered highly influential in the self-management of CFD. The MAGIC programme is an educational resource designed to facilitate self-management and its primary focus was to address these identified deficits, to enable people with CFD to develop self-management skills to effectively manage their CFD. The MAGIC programme was developed as a staged approach, this facilitates learning by building upon skills and knowledge previously learnt and developed. One of the aims of this approach is to try to improve an individual’s self-

efficacy in managing their CFD. The focus of the MAGIC programme was therefore on the physical needs of people with CFD. Further research on the emotional needs of people with CFD is required to inform appropriate healthcare pathways.

The MAGIC programme is a unique web-based resource, highly regarded by the people with CFD involved in the study. Its major benefits include: its strong-evidence base, meeting NICE criteria, web-based format which allows for more timely delivery and its relevance to service users as it is a resource designed for, and by, the people it was intended for. Further research to continue this work includes a feasibility study to assess the efficacy of the MAGIC programme in the 'real-world' setting and to develop a core outcome set relevant to CFD. This will then inform a rare disease trial required to demonstrate the effectiveness and cost effectiveness of the MAGIC programme so it can be incorporated into routine CF care and be of benefit to everyone with CFD.

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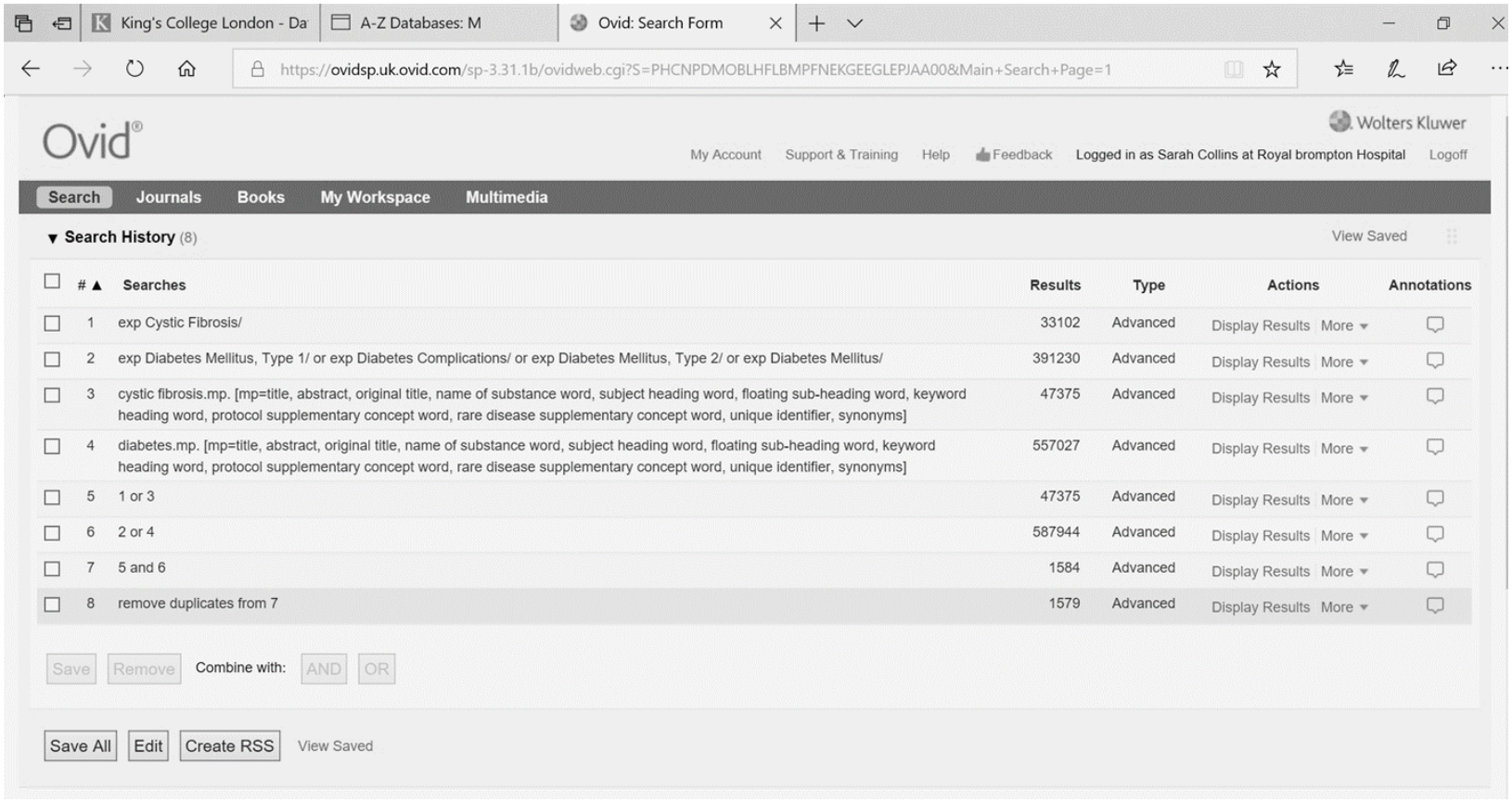
# Appendices

## Appendix 1: Training programme

| Title | Start Date | Internal/ External | Duration |
|---|---|---|---|
| Search Techniques for Systematic Reviews | 10-Aug-16 | Internal | 0.5 day |
| MicroSoft Office - long documents for theses | 16-Aug-16 | External | 1.0 day |
| HRA Research Day | 09-Sep-16 | External | 1 day |
| Managing your research data | 13-Sep-16 | Internal | 0.5 day |
| Advanced Quantitative Research Methods Module (Level 7- 30 credits) | 17-Sep-16 | Internal | 1 term |
| Starting Your PhD in the Sciences (For Postgraduate Research Students) | 31-Oct-16 | Internal | 0.5 day |
| Microsoft Excel Introduction | 07-Nov-16 | External | 1 day |
| RSM Royal Society of Medicine- Cystic fibrosis | 15-Nov-16 | External | 1 day |
| Desktop EndNote for PC | 22-Nov-16 | Internal | 0.5 day |
| Preparing for the Upgrade from MPhil to PhD in the Arts, Humanities & Social Sciences (For Postgraduate Research Students) | 07-Dec-16 | Internal | 0.5 day |
| Insulin pump course | 08-Dec-16 | External | 2 days |
| Advanced Qualitative Research Methods Module (Level 7- 30 credits) | 09-Jan-17 | Internal | 1 term |
| HEE/NIHR ICA meeting | 02-Feb-17 | External | 1 day |
| NIHR mentor winter school | 06-Feb-17 | External | 1 day |
| Diabetes UK Conference | 08-Mar-17 | External | 2 days |

| | | | |
|---|---|---|---|
| Clear & Concise Academic Writing (For Postgraduate Research Students & Research Staff) | 15-Mar-17 | Internal | 0.5 day |
| Microsoft Excel Intermediate | 07-Apr-17 | External | 1 day |
| Cystic Fibrosis Diabetes meeting | 03-May-17 | External | 1 day |
| CF Dietitians' Group meeting | 18-May-17 | External | 1 day |
| European CF Conference | 08-Jun-17 | External | 3 days |
| Microsoft Excel advanced | 22-Jun-17 | External | 1 day |
| working in clinical research network- CRN | 11-Jul-17 | External | 0.5 days |
| Qualitative Summer School- IPA masterclass | 17-Jul-17 | External | 1 day |
| IPA workshop- Derby | 19-Jul-17 | External | 1 day |
| GTA - Preparing to Teach in Higher Education for GTAs | 13-Sep-17 | External | 1 day |
| CF dietitians group meeting | 14-Nov-17 | External | 1 day |
| PhD education day | 21-Nov-17 | Internal | 1 day |
| PowerPoint for posters | 04-Dec-17 | External | 1 day |
| Level 3 award in Education and Training | 06-Dec-17 | External | 3 days |
| Cognitive Interview skills | 16-Jan-18 | External | 1 day |
| Introduction to data visualisation and infographics | 15-Mar-18 | External | 1 day |
| Supplementary Prescribing (Level 7 - 30 credits) | 22-Mar-18 | External | 6 months |
| European CF Conference | 05-Jun-18 | External | 5 days |
| CATO research symposium | 27-Jun-18 | External | 0.5 day |
| IPA meeting | 10-Jul-18 | External | 0.3 days |
| Springboard Women's Development Programme (For Research Staff & Postgraduate Research Students) | 09-Nov-18 | External | 4 days |
| CF dietitian's Group meeting | 13-Nov-13 | External | 1 day |
| IPA meeting | 10-Jan-19 | External | 0.3 days |
| NIHR conference | 07-Feb-19 | External | 1 day |

# Appendix 2: OVID Medline search strategy



King's College London - Da | A-Z Databases: M | Ovid: Search Form

https://ovidsp.uk.ovid.com/sp-3.31.1b/ovidweb.cgi?S=PHCNPDMOBLHFLBMPFNEKGEEGLEPJAA00&Main+Search+Page=1

Ovid® — Wolters Kluwer

My Account | Support & Training | Help | Feedback | Logged in as Sarah Collins at Royal brompton Hospital | Logoff

Search | Journals | Books | My Workspace | Multimedia

▼ Search History (8) — View Saved

| # ▲ | Searches | Results | Type | Actions | Annotations |
|---|---|---|---|---|---|
| 1 | exp Cystic Fibrosis/ | 33102 | Advanced | Display Results \| More | |
| 2 | exp Diabetes Mellitus, Type 1/ or exp Diabetes Complications/ or exp Diabetes Mellitus, Type 2/ or exp Diabetes Mellitus/ | 391230 | Advanced | Display Results \| More | |
| 3 | cystic fibrosis.mp. [mp=title, abstract, original title, name of substance word, subject heading word, floating sub-heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier, synonyms] | 47375 | Advanced | Display Results \| More | |
| 4 | diabetes.mp. [mp=title, abstract, original title, name of substance word, subject heading word, floating sub-heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier, synonyms] | 557027 | Advanced | Display Results \| More | |
| 5 | 1 or 3 | 47375 | Advanced | Display Results \| More | |
| 6 | 2 or 4 | 587944 | Advanced | Display Results \| More | |
| 7 | 5 and 6 | 1584 | Advanced | Display Results \| More | |
| 8 | remove duplicates from 7 | 1579 | Advanced | Display Results \| More | |

Save | Remove | Combine with: | AND | OR

Save All | Edit | Create RSS | View Saved

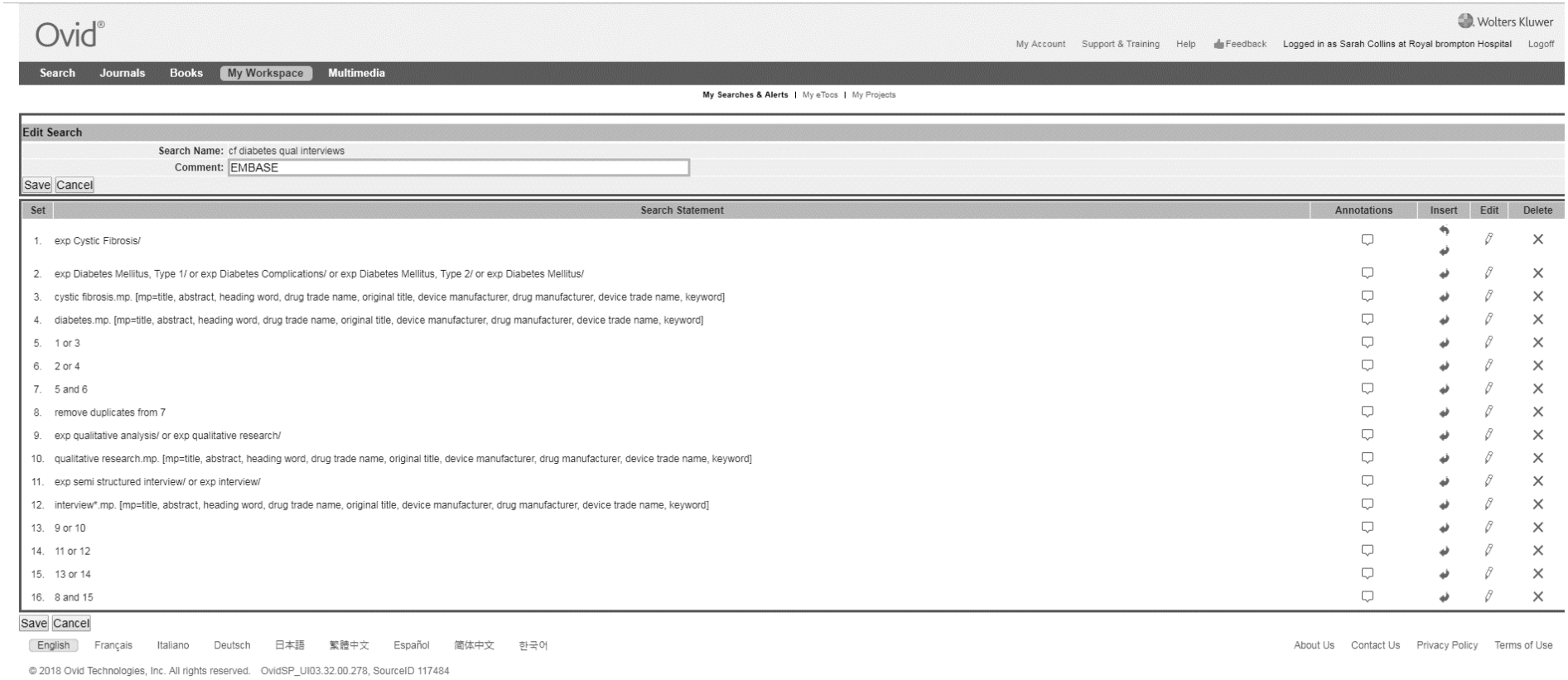
# Appendix 3: CINAHL search strategy

Save Cancel

Search History

| # | Query | Limiters/Expanders | Last Run Via |
|---|---|---|---|
| S16 | S7 AND S15 | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S15 | S13 OR S14 | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S14 | S11 OR S12 | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S13 | S8 OR S9 OR S10 | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S12 | interviews | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S11 | (MH "Interviews+") | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S10 | qualitative research | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S9 | qualitative studies | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S8 | (MH "Qualitative Studies") | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S7 | S5 AND S6 | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S6 | S3 OR S4 | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S5 | S1 OR S2 | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S4 | diabetes | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S3 | (MH "Diabetes Mellitus, Type 1") OR (MH "Diabetes Mellitus, Type 2") OR (MH "Diabetes Mellitus") | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S2 | cystic fibrosis | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search<br>Database - CINAHL |
| S1 | (MH "Cystic Fibrosis") | Search modes - Boolean/Phrase | Interface - EBSCOhost Research Databases<br>Search Screen - Advanced Search |

# Appendix 4: EMBASE search strategy



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My Account | Support & Training | Help | Feedback | Logged in as Sarah Collins at Royal brompton Hospital | Logoff

Search | Journals | Books | My Workspace | Multimedia

My Searches & Alerts | My eTocs | My Projects

**Edit Search**

Search Name: cf diabetes qual interviews

Comment: EMBASE

Save Cancel

| Set | Search Statement | Annotations | Insert | Edit | Delete |
|---|---|---|---|---|---|
| 1. | exp Cystic Fibrosis/ | | | | |
| 2. | exp Diabetes Mellitus, Type 1/ or exp Diabetes Complications/ or exp Diabetes Mellitus, Type 2/ or exp Diabetes Mellitus/ | | | | |
| 3. | cystic fibrosis.mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword] | | | | |
| 4. | diabetes.mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword] | | | | |
| 5. | 1 or 3 | | | | |
| 6. | 2 or 4 | | | | |
| 7. | 5 and 6 | | | | |
| 8. | remove duplicates from 7 | | | | |
| 9. | exp qualitative analysis/ or exp qualitative research/ | | | | |
| 10. | qualitative research.mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword] | | | | |
| 11. | exp semi structured interview/ or exp interview/ | | | | |
| 12. | interview*.mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword] | | | | |
| 13. | 9 or 10 | | | | |
| 14. | 11 or 12 | | | | |
| 15. | 13 or 14 | | | | |
| 16. | 8 and 15 | | | | |

Save Cancel

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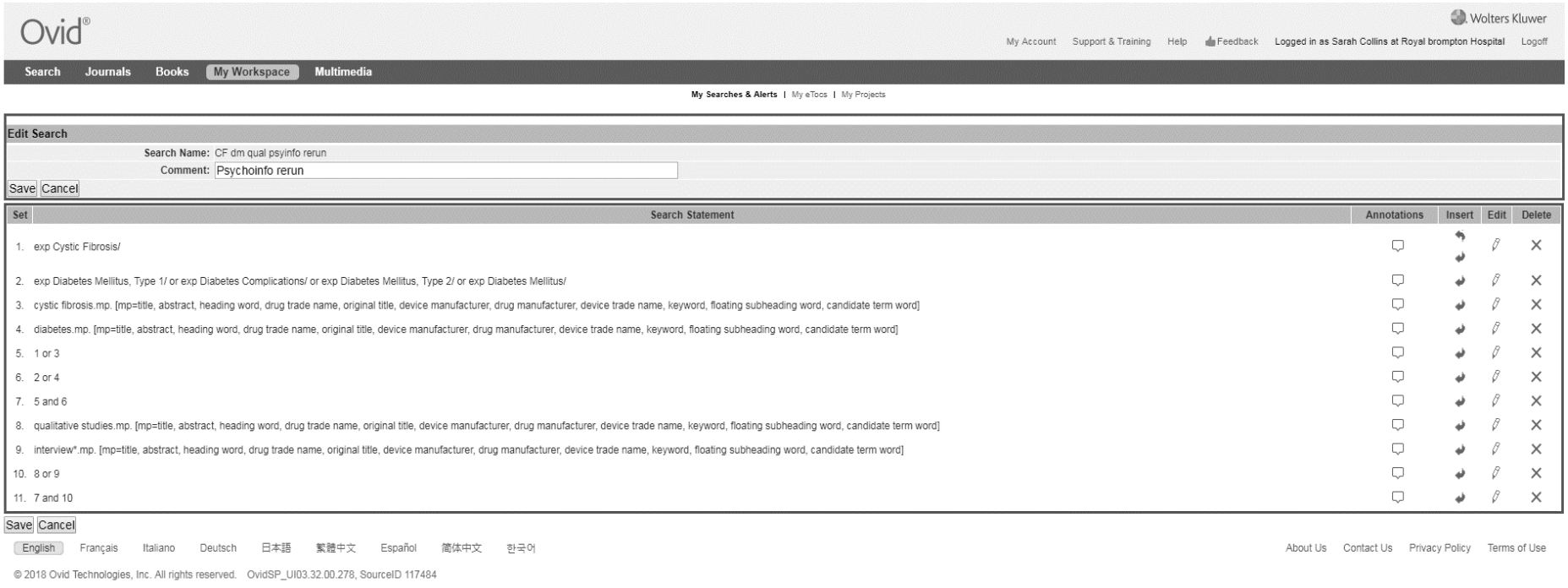
# Appendix 5: PsychInfo search strategy



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Search | Journals | Books | My Workspace | Multimedia

My Searches & Alerts | My eToos | My Projects

**Edit Search**

**Search Name:** CF dm qual psyinfo rerun

**Comment:** Psychoinfo rerun

Save Cancel

| Set | Search Statement | Annotations | Insert | Edit | Delete |
|---|---|---|---|---|---|
| 1. | exp Cystic Fibrosis/ | | | | |
| 2. | exp Diabetes Mellitus, Type 1/ or exp Diabetes Complications/ or exp Diabetes Mellitus, Type 2/ or exp Diabetes Mellitus/ | | | | |
| 3. | cystic fibrosis.mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword, floating subheading word, candidate term word] | | | | |
| 4. | diabetes.mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword, floating subheading word, candidate term word] | | | | |
| 5. | 1 or 3 | | | | |
| 6. | 2 or 4 | | | | |
| 7. | 5 and 6 | | | | |
| 8. | qualitative studies.mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword, floating subheading word, candidate term word] | | | | |
| 9. | interview*.mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword, floating subheading word, candidate term word] | | | | |
| 10. | 8 or 9 | | | | |
| 11. | 7 and 10 | | | | |

Save Cancel

English | Français | Italiano | Deutsch | 日本語 | 繁體中文 | Español | 简体中文 | 한국어

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# Appendix 6: Meta-ethnography reporting guidelines

Application of the reporting criteria to this study

| No. | Criteria Headings | Reporting Criteria | Comments- in relation to this study |
|---|---|---|---|
| **Phase 1—Selecting meta-ethnography and getting started** | | | |
| ***Introduction*** | | | |
| **1** | Rationale and context for the meta-ethnography | Describe the gap in research or knowledge to be filled by the meta-ethnography, and the wider context of the meta-ethnography | In methodology chapter |
| **2** | Aim(s) of the meta-ethnography | Describe the meta-ethnography aim(s) | In methodology chapter |
| **3** | Focus of the meta-ethnography | Describe the meta-ethnography review question(s) (or objectives) | In methodology chapter |
| **4** | Rationale for using meta-ethnography | Explain why meta-ethnography was considered the most appropriate qualitative synthesis methodology | In methodology chapter |
| **Phase 2—Deciding what is relevant** | | | |
| ***Methods*** | | | |
| **5** | Search strategy | Describe the rationale for the literature search strategy | completed |
| **6** | Search processes | Describe how the literature searching was carried out and by whom | completed |
| **7** | Selecting primary studies | Describe the process of study screening and selection, and who was involved | completed |
| ***Findings*** | | | |
| **8** | Outcome of study selection | Describe the results of study searches and screening | Presented as flowchart |
| **Phase 3—Reading included studies** | | | |
| ***Methods*** | | | |
| **9** | Reading and data extraction approach | Describe the reading and data extraction method and processes | completed |
| ***Findings*** | | | |
| **10** | Presenting characteristics of included studies | Describe characteristics of the included studies | Characteristics presented as a table |
| **Phase 4—Determining how studies are related** | | | |
| ***Methods*** | | | |
| **11** | Process for determining how studies are related | Describe the methods and processes for determining how the included studies are related: | Yes, brief summary of similarities only. |

| | | | |
|---|---|---|---|
| | | - Which aspects of studies were compared<br><br>AND<br><br>- How the studies were compared | |
| ***Findings*** | | | |
| **12** | Outcome of relating studies | Describe how studies relate to each other | Brief |
| **Phase 5—Translating studies into one another** | | | |
| ***Methods*** | | | |
| **13** | Process of translating studies | Describe the methods of translation**:**<br><br>- Describe steps taken to preserve the context and meaning of the relationships between concepts within and across studies- Describe how the reciprocal and refutational translations were conducted- Describe how potential alternative interpretations or explanations were considered in the translations | Discussed. Use of supervisory group to explore interpretations. |
| ***Findings*** | | | |
| **14** | Outcome of translation | Describe the interpretive findings of the translation. | Presented as table of occurrence of themes |
| **Phase 6—Synthesizing translations** | | | |
| ***Methods*** | | | |
| **15** | Synthesis process | Describe the methods used to develop overarching concepts ("synthesised translations"). Describe how potential alternative interpretations or explanations were considered in the synthesis | Discussed |
| ***Findings*** | | | |
| **16** | Outcome of synthesis process | Describe the new theory, conceptual framework, model, configuration, or interpretation of data developed from the synthesis | Conceptual model developed |
| **Phase 7—Expressing the synthesis** | | | |
| ***Discussion*** | | | |
| **17** | Summary of findings | Summarise the main interpretive findings of the translation and synthesis and compare them to existing literature | completed |
| **18** | Strengths, limitations, and reflexivity | Reflect on and describe the strengths and limitations of the synthesis: | completed |

| | | | |
|---|---|---|---|
| | | - Methodological aspects—for example, describe how the synthesis findings were influenced by the nature of the included studies and how the meta-ethnography was conducted. - Reflexivity—for example, the impact of the research team on the synthesis findings | |
| **19** | Recommendations and conclusions | Describe the implications of the synthesis | completed |

# Appendix 7: REC favourable opinion

NHS
Health Research Authority

London - South East Research Ethics Committee
Barlow House
3rd Floor
4 Minshull Street
Manchester
M1 3DZ

Telephone: 0161 625 7109
Fax: 0161 625 7919

**Please note: This is the favourable opinion of the REC only and does not allow you to start your study at NHS sites in England until you receive HRA Approval**

14 March 2017

Miss Sarah Collins
HEE/NIHR Clinical Doctoral Research Fellow
Royal Brompton and Harefield NHS Trust
Cystic Fibrosis Department
Royal Brompton Hospital
London
SW3 6NP

Dear Miss Collins

| | |
|---|---|
| **Study title:** | **Managing abnormal glucose in cystic fibrosis (MAGIC)- the development of a self-management education programme for people with CFRD** |
| **REC reference:** | **17/LO/0377** |
| **IRAS project ID:** | **215946** |

The Proportionate Review Sub-committee of the London - South East Research Ethics Committee reviewed the above application on 23 February 2017.

We plan to publish your research summary wording for the above study on the HRA website, together with your contact details. Publication will be no earlier than three months from the date of this favourable opinion letter. The expectation is that this information will be published for all studies that receive an ethical opinion but should you wish to provide a substitute contact point, wish to make a request to defer, or require further information, please contact hra.studyregistration@nhs.net outlining the reasons for your request. Under very limited circumstances (e.g. for student research which has received an unfavourable opinion), it may be possible to grant an exemption to the publication of the study.

**Ethical opinion**

On behalf of the Committee, the sub-committee gave a favourable ethical opinion of the above research on the basis described in the application form, protocol and supporting documentation, subject to the conditions specified below.

**Conditions of the favourable opinion**

The REC favourable opinion is subject to the following conditions being met prior to the start of the study.

Management permission must be obtained from each host organisation prior to the start of the study at the site concerned.

*Management permission should be sought from all NHS organisations involved in the study in accordance with NHS research governance arrangements. Each NHS organisation must confirm through the signing of agreements and/or other documents that it has given permission for the research to proceed (except where explicitly specified otherwise).*

*Guidance on applying for HRA Approval (England)/ NHS permission for research is available in the Integrated Research Application System, www.hra.nhs.uk or at http://www.rdforum.nhs.uk.*

*Where a NHS organisation's role in the study is limited to identifying and referring potential participants to research sites ("participant identification centre"), guidance should be sought from the R&D office on the information it requires to give permission for this activity.*

*For non-NHS sites, site management permission should be obtained in accordance with the procedures of the relevant host organisation.*

*Sponsors are not required to notify the Committee of management permissions from host organisations.*

Registration of Clinical Trials

All clinical trials (defined as the first four categories on the IRAS filter page) must be registered on a publically accessible database. This should be before the first participant is recruited but no later than 6 weeks after recruitment of the first participant.

There is no requirement to separately notify the REC but you should do so at the earliest opportunity e.g. when submitting an amendment. We will audit the registration details as part of the annual progress reporting process.

To ensure transparency in research, we strongly recommend that all research is registered but for non-clinical trials this is not currently mandatory.

If a sponsor wishes to request a deferral for study registration within the required timeframe, they should contact hra.studyregistration@nhs.net. The expectation is that all clinical trials will be registered, however, in exceptional circumstances non registration may be permissible with prior agreement from the HRA. Guidance on where to register is provided on the HRA website.

**It is the responsibility of the sponsor to ensure that all the conditions are complied with before the start of the study or its initiation at a particular site (as applicable).**

**Ethical review of research sites**

The favourable opinion applies to all NHS sites taking part in the study, subject to management permission being obtained from the NHS/HSC R&D office prior to the start of the study (see "Conditions of the favourable opinion").

**Summary of discussion at the meeting**

The PR Sub Committee raised no Ethical Issues with this Application and was content to issue a Favourable Opinion.

**Approved documents**

The documents reviewed and approved were:

| *Document* | *Version* | *Date* |
|---|---|---|
| Interview schedules or topic guides for participants | | 02 February 2017 |
| Interview schedules or topic guides for participants [Participant Interview Schedule] | 1 | 01 February 2017 |
| IRAS Application Form [IRAS_Form_09022017] | | 09 February 2017 |
| IRAS Checklist XML [Checklist_09022017] | | 09 February 2017 |
| Letters of invitation to participant [Qualitative Interview ] | 1.0 | 01 November 2016 |
| Letters of invitation to participant [MAGIC Evaluation] | 1.0 | 01 November 2016 |
| Participant consent form [Updated consent form] | 1 | 01 February 2017 |
| Participant information sheet (PIS) [PIS Magic Programme] | 1 | 03 February 2017 |
| Participant information sheet (PIS) [PIS Qualitative Interviews] | 1 | 03 February 2017 |
| Research protocol or project proposal | 1.2 | 03 February 2017 |
| Response to Request for Further Information [Addendum to the IRAS Form] | 2 | |
| Summary CV for Chief Investigator (CI) | | 01 February 2017 |
| Summary CV for supervisor (student research) [Jacqueline Alys Sturt ] | | |
| Summary CV for supervisor (student research) [Sue Woodward] | | 01 February 2017 |
| Summary CV for supervisor (student research) [Andrew Lee Jones] | | 03 February 2017 |

**Membership of the Proportionate Review Sub-Committee**

The members of the Sub-Committee who took part in the review are listed on the attached sheet.

**Statement of compliance**

The Committee is constituted in accordance with the Governance Arrangements for Research Ethics Committees and complies fully with the Standard Operating Procedures for Research Ethics Committees in the UK.

**After ethical review**

Reporting requirements

The attached document "After ethical review – guidance for researchers" gives detailed guidance on reporting requirements for studies with a favourable opinion, including:

- Notifying substantial amendments
- Adding new sites and investigators
- Notification of serious breaches of the protocol
- Progress and safety reports
- Notifying the end of the study

The HRA website also provides guidance on these topics, which is updated in the light of changes in reporting requirements or procedures.

**User Feedback**

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and

the application procedure. If you wish to make your views known please use the feedback form available on the HRA website: http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/

**HRA Training**

We are pleased to welcome researchers and R&D staff at our training days – see details at http://www.hra.nhs.uk/hra-training/

With the Committee's best wishes for the success of this project.

| | |
|---|---|
| **17/LO/0377** | **Please quote this number on all correspondence** |

Yours sincerely

PP
**Professor David Caplin**
**Chair**

Email: nrescommittee.london-southeast@nhs.net

*Enclosures:* *List of names and professions of members who took part in the review*

*"After ethical review – guidance for researchers"*

*Copy to:* *Mr Patrik Pettersson, Royal Brompton & Harefield Foundation Trust*

**London - South East Research Ethics Committee**

**Attendance at PRS Sub-Committee of the REC meeting on 23 February 2017**

**Committee Members:**

| *Name* | *Profession* | *Present* | *Notes* |
|---|---|---|---|
| Professor David Caplin | Physicist | Yes | Chair |
| Ms Janelle Hill | Former Banking Administrator | Yes | |
| Ms Vanda Taylor | Senior Cancer Information Nurse | Yes | |

# Appendix 8: HRA approval

NHS
Health Research Authority

Miss Sarah Collins
HEE/NIHR Clinical Doctoral Research Fellow
Royal Brompton and Harefield NHS Trust
Cystic Fibrosis Department
Royal Brompton Hospital
London
SW3 6NP

Email: hra.approval@nhs.net

30 March 2017

Dear Miss Collins

**Letter of HRA Approval**

| | |
|---|---|
| **Study title:** | **Managing abnormal glucose in cystic fibrosis (MAGIC)- the development of a self-management education programme for people with CFRD** |
| **IRAS project ID:** | **215946** |
| **REC reference:** | **17/LO/0377** |
| **Sponsor** | **Royal Brompton & Harefield Foundation Trust** |

I am pleased to confirm that HRA Approval has been given for the above referenced study, on the basis described in the application form, protocol, supporting documentation and any clarifications noted in this letter.

**Participation of NHS Organisations in England**

The sponsor should now provide a copy of this letter to all participating NHS organisations in England.

*Appendix B* provides important information for sponsors and participating NHS organisations in England for arranging and confirming capacity and capability. **Please read *Appendix B* carefully**, in particular the following sections:

- *Participating NHS organisations in England* – this clarifies the types of participating organisations in the study and whether or not all organisations will be undertaking the same activities
- *Confirmation of capacity and capability* - this confirms whether or not each type of participating NHS organisation in England is expected to give formal confirmation of capacity and capability. Where formal confirmation is not expected, the section also provides details on the time limit given to participating organisations to opt out of the study, or request additional time, before their participation is assumed.
- *Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria)* - this provides detail on the form of agreement to be used in the study to confirm capacity and capability, where applicable.

Further information on funding, HR processes, and compliance with HRA criteria and standards is also provided.

It is critical that you involve both the research management function (e.g. R&D office) supporting each organisation and the local research team (where there is one) in setting up your study. Contact details and further information about working with the research management function for each organisation can be accessed from www.hra.nhs.uk/hra-approval.

**Appendices**

The HRA Approval letter contains the following appendices:

- A – List of documents reviewed during HRA assessment
- B – Summary of HRA assessment

**After HRA Approval**

The document *"After Ethical Review – guidance for sponsors and investigators"*, issued with your REC favourable opinion, gives detailed guidance on reporting expectations for studies, including:

- Registration of research
- Notifying amendments
- Notifying the end of the study

The HRA website also provides guidance on these topics, and is updated in the light of changes in reporting expectations or procedures.

In addition to the guidance in the above, please note the following:

- HRA Approval applies for the duration of your REC favourable opinion, unless otherwise notified in writing by the HRA.
- Substantial amendments should be submitted directly to the Research Ethics Committee, as detailed in the *After Ethical Review* document. Non-substantial amendments should be submitted for review by the HRA using the form provided on the HRA website, and emailed to hra.amendments@nhs.net.
- The HRA will categorise amendments (substantial and non-substantial) and issue confirmation of continued HRA Approval. Further details can be found on the HRA website.

**Scope**

HRA Approval provides an approval for research involving patients or staff in NHS organisations in England.

If your study involves NHS organisations in other countries in the UK, please contact the relevant national coordinating functions for support and advice. Further information can be found at http://www.hra.nhs.uk/resources/applying-for-reviews/nhs-hsc-rd-review/.

If there are participating non-NHS organisations, local agreement should be obtained in accordance with the procedures of the local participating non-NHS organisation.

| IRAS project ID | 215946 |
|---|---|

**User Feedback**

The Health Research Authority is continually striving to provide a high quality service to all applicants and sponsors. You are invited to give your view of the service you have received and the application procedure. If you wish to make your views known please use the feedback form available on the HRA website: http://www.hra.nhs.uk/about-the-hra/governance/quality-assurance/.

**HRA Training**

We are pleased to welcome researchers and research management staff at our training days – see details at http://www.hra.nhs.uk/hra-training/

Your IRAS project ID is **215946**. Please quote this on all correspondence.

Yours sincerely

Joanna Ho
Assessor

Email: hra.approval@nhs.net

*Copy to:* *Mr Patrik Pettersson, Sponsor Representative and Lead NHS R&D Contact, Royal Brompton & Harefield Foundation Trust*
*Professor Jackie Sturt, Academic Supervisor, King's College London*

| IRAS project ID | 215946 |
|---|---|

## Appendix A - List of Documents

The final document set assessed and approved by HRA Approval is listed below.

| *Document* | *Version* | *Date* |
|---|---|---|
| Interview schedules or topic guides for participants | | 02 February 2017 |
| Interview schedules or topic guides for participants [Participant Interview Schedule] | 1 | 01 February 2017 |
| IRAS Application Form [IRAS_Form_09022017] | | 09 February 2017 |
| IRAS Checklist XML [Checklist_09022017] | | 09 February 2017 |
| Letter from funder [Funding letter] | 1.0 | 25 February 2016 |
| Letters of invitation to participant [Qualitative Interview ] | 1.0 | 01 November 2016 |
| Letters of invitation to participant [MAGIC Evaluation] | 1.0 | 01 November 2016 |
| Other [NIHR Approval] | 1.0 | 16 February 2016 |
| Other [Signed Contract Page] | 1.0 | 27 July 2016 |
| Other [Payment Schedule] | 1.0 | 25 January 2015 |
| Participant consent form [Updated consent form] | 1 | 01 February 2017 |
| Participant information sheet (PIS) [PIS Magic Programme] | 1 | 03 February 2017 |
| Participant information sheet (PIS) [PIS Qualitative Interviews] | 1 | 03 February 2017 |
| Research protocol or project proposal | 1.2 | 03 February 2017 |
| Response to Request for Further Information [Addendum to the IRAS Form] | 2 | |
| Summary CV for Chief Investigator (CI) | | 01 February 2017 |
| Summary CV for supervisor (student research) [Jacqueline Alys Sturt ] | | |
| Summary CV for supervisor (student research) [Sue Woodward] | | 01 February 2017 |
| Summary CV for supervisor (student research) [Andrew Lee Jones] | | 03 February 2017 |

| IRAS project ID | 215946 |
|---|---|

**Appendix B - Summary of HRA Assessment**

This appendix provides assurance to you, the sponsor and the NHS in England that the study, as reviewed for HRA Approval, is compliant with relevant standards. It also provides information and clarification, where appropriate, to participating NHS organisations in England to assist in assessing and arranging capacity and capability.

**For information on how the sponsor should be working with participating NHS organisations in England, please refer to the, *participating NHS organisations, capacity and capability* and *Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria)* sections in this appendix.**

The following person is the sponsor contact for the purpose of addressing participating organisation questions relating to the study:

Name: Mr Patrik Pettersson
Tel: 0207 351 8736
Email: p.pettersson@rbht.nhs.uk

## HRA assessment criteria

| Section | HRA Assessment Criteria | Compliant with Standards | Comments |
|---|---|---|---|
| 1.1 | IRAS application completed correctly | Yes | No comments |
| | | | |
| 2.1 | Participant information/consent documents and consent process | Yes | It was noted that the IRAS reference was not included in the participant information and consent documents. Sponsor should consider to include this in any subsequent amendment so that participants have a single reference for the study. |
| | | | |
| 3.1 | Protocol assessment | Yes | No comments |
| | | | |
| 4.1 | Allocation of responsibilities and rights are agreed and documented | Yes | No agreement is expected as the participating NHS organisation is also the study sponsor. |
| 4.2 | Insurance/indemnity arrangements assessed | Yes | NHS indemnity applies to the management, design and conduct of the study. |

| Section | HRA Assessment Criteria | Compliant with Standards | Comments |
|---|---|---|---|
| | | | Where applicable, independent contractors (e.g. General Practitioners) should ensure that the professional indemnity provided by their medical defence organisation covers the activities expected of them for this research study |
| 4.3 | Financial arrangements assessed | Yes | Funding has been secured from NIHR for this study. |
| | | | |
| 5.1 | Compliance with the Data Protection Act and data security issues assessed | Yes | No comments |
| 5.2 | CTIMPS – Arrangements for compliance with the Clinical Trials Regulations assessed | Not Applicable | No comments |
| 5.3 | Compliance with any applicable laws or regulations | Yes | No comments |
| | | | |
| 6.1 | NHS Research Ethics Committee favourable opinion received for applicable studies | Yes | REC Favourable Opinion issued 14 March 2017 |
| 6.2 | CTIMPS – Clinical Trials Authorisation (CTA) letter received | Not Applicable | No comments |
| 6.3 | Devices – MHRA notice of no objection received | Not Applicable | No comments |
| 6.4 | Other regulatory approvals and authorisations received | Not Applicable | No comments |

## Participating NHS Organisations in England

| *This provides detail on the types of participating NHS organisations in the study and a statement as to whether the activities at all organisations are the same or different.* |
|---|
| There is participating NHS organisation in this study which is also the study sponsor. All study activities as detailed in the study documents will take place at site.<br><br>If this study is subsequently extended to other NHS organisation(s) in England, an amendment |

should be submitted to the HRA, with a Statement of Activities and Schedule of Events for the newly participating NHS organisation(s) in England.

The Chief Investigator or sponsor should share relevant study documents with participating NHS organisations in England in order to put arrangements in place to deliver the study. The documents should be sent to both the local study team, where applicable, and the office providing the research management function at the participating organisation. For NIHR CRN Portfolio studies, the Local LCRN contact should also be copied into this correspondence. For further guidance on working with participating NHS organisations please see the HRA website.

If chief investigators, sponsors or principal investigators are asked to complete site level forms for participating NHS organisations in England which are not provided in IRAS or on the HRA website, the chief investigator, sponsor or principal investigator should notify the HRA immediately at hra.approval@nhs.net. The HRA will work with these organisations to achieve a consistent approach to information provision.

## Confirmation of Capacity and Capability

*This describes whether formal confirmation of capacity and capability is expected from participating NHS organisations in England.*

The HRA has determined that participating NHS organisations in England **are not expected to formally confirm their capacity and capability to host this research,** because the participating site is also the study sponsor.

- The HRA has informed the relevant research management offices that you intend to undertake the research at their organisation. However, you should still support and liaise with these organisations as necessary.
- Following issue of the Letter of HRA Approval the sponsor may commence the study at these organisations when it is ready to do so.
- The document "Collaborative working between sponsors and NHS organisations in England for HRA Approval studies, where no formal confirmation of capacity and capability is expected" provides further information for the sponsor and NHS organisations on working with NHS organisations in England where no formal confirmation of capacity and capability is expected, and the processes involved in adding new organisations. Further study specific details are provided the *Participating NHS Organisations* and *Allocation of responsibilities and rights are agreed and documented (4.1 of HRA assessment criteria)* sections of this Appendix.

## Principal Investigator Suitability

*This confirms whether the sponsor position on whether a PI, LC or neither should be in place is correct for each type of participating NHS organisation in England and the minimum expectations for education, training and experience that PIs should meet (where applicable).*

A Principal Investigator has been identified for the participating NHS organisation.

GCP training is not a generic training expectation, in line with the HRA statement on training

expectations.

### HR Good Practice Resource Pack Expectations

| *This confirms the HR Good Practice Resource Pack expectations for the study and the pre-engagement checks that should and should not be undertaken* |
|---|
| Local staff who are substantively employed by the participating NHS organisation will be undertaking all study related activities as described in the research application. Therefore, no further HR arrangements are expected for this study. |

### Other Information to Aid Study Set-up

| *This details any other information that may be helpful to sponsors and participating NHS organisations in England to aid study set-up.* |
|---|
| The applicant has indicated that they intend to apply for inclusion on the NIHR CRN Portfolio. |

# Appendix 9: Royal Brompton and Harefield NHS foundation Trust approval


Royal Brompton & Harefield NHS
NHS Foundation Trust

**Research Office**
**Royal Brompton Hospital**
**Chelsea Wing, Level 2**
**Sydney Street**
**SW3 6NP**
www.rbht.nhs.uk

05 May 2017

Miss Sarah Collins
HEE/NIHR Clinical Doctoral Research Fellow
Royal Brompton and Harefield NHS Trust
Cystic Fibrosis Department
Royal Brompton Hospital

Dear Sarah

| | |
|---|---|
| **Project Title:** | Managing abnormal glucose in cystic fibrosis (MAGIC) - the development of a self-management education programme for people with CFRD |
| **REC Ref:** | 17/LO/0377 |
| **IRAS Project ID:** | 215946 |
| **Study Sponsor:** | Royal Brompton & Harefield NHS Foundation Trust |

| | |
|---|---|
| **Recruitment End Date:** | **30/12/2018** |
| **Study End Date:** | **31/05/2019** |
| **Recruitment target:** | **20** |

## Confirmation of Capacity and Capability

Thank you for registering the above research project with the Research Office.

I am pleased to provide confirmation that the Trust has 'capacity' to participate and 'capability' to deliver the above project based on the arrangements outlined in the Statement of Activity as outlined in the Health Research Authority (HRA) approval.

This Confirmation of Capacity and Capability is granted on the basis that it will be conducted as described in the study protocol and supporting documentation as approved by the HRA, and on the understanding that the study is conducted in accordance with the principles set out in the Research Governance Framework for Health and Social Care (April 2005, 2nd Edition) and RB&HFT Policies and procedures.

### Patient recruitment

The Trust is contractually obliged to measure and publish data on the days elapsing between the time we receive a valid research application and the time when the first participant is recruited to the trial. This applies to all clinical research where confirmation of capacity and capability has been issued.

The Trust is obliged to regularly report on various research metrics to the National Institute of Health and Research (NIHR) and Department of Health (DoH).

It is the responsibility of the local Principal Investigator (PI) to ensure that the following responsibilities are appropriately delegated within the project research team to ensure timely communication with the Research Office:

- The 1st project participant is recruited within 30 days of issue of this letter,
- That all participant recruitment is recorded, on a monthly basis,
- That monthly recruitment numbers (accruals) are uploaded to the NIHR Central Portfolio Management System (CPMS) if you are the study Chief Investigator (CI). Alternatively the PI should ensure that this information is notified to the CI.

**Study Amendments**

HRA approval applies for the duration of the REC favourable opinion, unless otherwise notified in writing by the HRA. It is the responsibility of the Sponsor to ensure that all study amendments are submitted to the HRA and notified to the Research Office and study research team in a timely manner.

Changes to the status of the project, including study suspension or premature termination, should also be communicated to the Trust Research Office.

**Safety Reporting**

The research Sponsor, the Chief Investigator (CI) or the local Principal Investigator (PI) at a research site, may take appropriate Urgent Safety Measures in order to protect research participants against any immediate hazard to their health or safety. The Research Office should be notified of such measures immediately. The notification should include reasons why the measures were taken and the plan for further action.

All patient related incidents must be reported internally by the study team in line with the Trust Adverse Incident Management and Reporting Policy *via* the Quality and Safety Department database **Datix** and marked "research-related".

In addition, **all** Serious Adverse Events/Reactions (SAE/Rs) must be reported to the study Sponsor by a member of the study team immediately and as specified in the study protocol and sponsors' Pharmacovigilance SOP.

Copies of **all** SUSAR reports should be sent to the Research Office immediately and in parallel to informing the study Sponsor.

It is the responsibility of the study Sponsor to ensure that Development Safety Update Reports (DSURs) and quarterly safety reports are sent to the Research Office in a timely manner.

**Audit**

Please note the Trust is required to monitor research to ensure compliance with the Research Governance Framework and other legal and regulatory requirements. This responsibility is delegated to the Research Office and will be achieved by random audit of active research projects across the Trust in accordance with the RB&HFT Audit SOP.

Yours sincerely

Dr Jenny Rivers
Associate Director of Research

# Appendix 10: Confidentiality Agreement

The Transcription Company UK
74 Reddicap Heath Road
Sutton Coldfield
West Midlands
B75 7EN

The Transcription Company UK
www.thetranscription.co.uk

Tel/Fax: 0121 681 8058

info@thetranscription.co.uk

**CONFIDENTIALITY AGREEMENT**

This confidentiality agreement has been prepared and is being distributed on behalf of The Transcription Company UK who acknowledges and accepts the terms and conditions of this Confidentiality Agreement.

The Transcription Company UK recognises that in the course of rendering transcription services, it will be exposed to and have in its possession Confidential Information. The Transcription Company UK pledges not to use any portion of the confidential information for any purpose other than that required for the fulfilment of transcription services for :

**Sarah Collins**
**Royal Brompton Hospital**
**Sydney Street**
**London**
**SW3 6NP**

Specifically, The Transcription Company UK will not at any time or in any manner, either directly or indirectly, use for personal gain, or divulge, disclose, or communicate in any manner and information that is proprietary to the Client, or otherwise divulge any portion of the Confidential Information to any third party without the prior written consent of the client.

The Transcription Company UK will protect such information and treat it as strictly confidential.

The Transcription Company UK acknowledges that the client reserves the right to terminate any contract should they become aware of any unauthorised use of the Confidential Information.

signed M A Cowley date: 10.7.2017
Melanie Cowley, Manager
The Transcription Company UK

# Appendix 11: Interview schedule

1. **Can you describe what it is like for you to live with CF?**
Possible prompts; typical day/ treatments/ feelings

2. **Can you tell me about when you were diagnosed with CFD?**
Possible prompts: thoughts/ feelings/ meanings/ treatments/coping/ awareness

3. **Can you describe the actual experience of living with CFD on a day-to-day basis?**
Possible prompts- thoughts/ feelings/ attitudes/ activities/ barriers/ difficulties/ what helps/ how fits in with CF/ impact on health/ what difficult or easy

4. **Can you describe how you learnt to manage your CFD?**
Possible prompts; support/ feelings/ resources

5. **Can you describe what it means to you having CFD?**
Prompt words/ images/ metaphors/ associations/ identity

6. **Please discuss how you feel you manage your CFD**
Possible prompts; barriers/ facilitators/ comparisons to CF/coping

7. **Can you discuss what it is like for you managing your CF and CFD?**
Possible prompts: conflicts/ challenges/ feelings/ identity

8. **Can you discuss how you see your future living with CFD?**
Possible prompts: compare and contrast with CF/ is one more important than the other

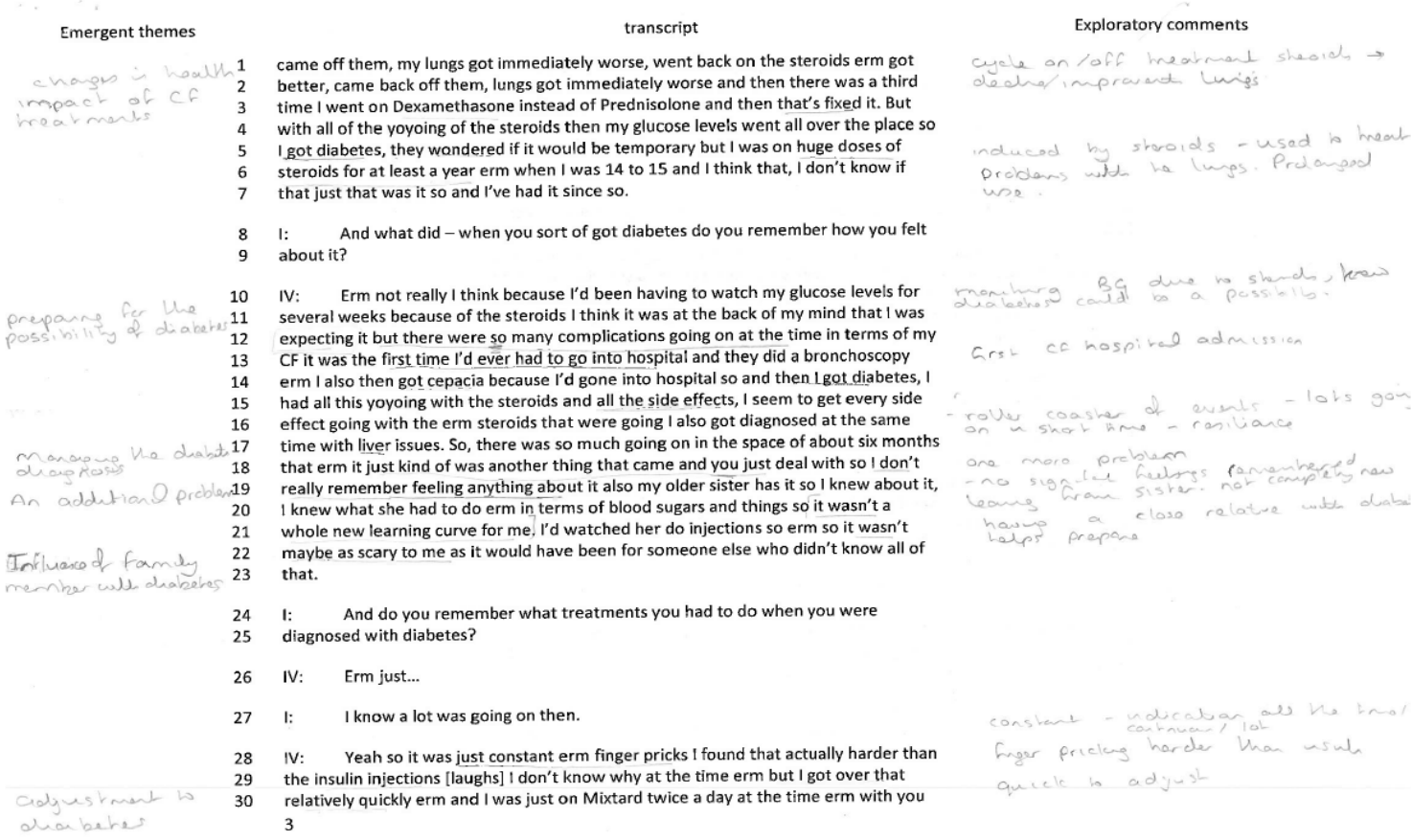
9. **Can you discuss any ideas that you have that will help people with CF manage their CFD?**
Possible prompts; types resources/ education/ support/what you think will help/ what doesn't help

10.**Is there anything else today that you would like to discuss about what it is like for you to**
**live with CFD?**

# Appendix 12: Application of key themes from meta-ethnography to interview topic guide

| | A need to develop a perspective on CFD | Struggling with identity | A representation of morbidity and mortality | In control or being controlled | Need for knowledge and support |
|---|---|---|---|---|---|
| Interview topic guide questions | Can you tell me about when you were diagnosed with CFD? | Can you tell me about when you were diagnosed with CFD? | | | |
| | Can you describe the actual experience of living with CFD on a day-to-day basis? | Can you describe the actual experience of living with CFD on a day-to-day basis? | Can you describe the actual experience of living with CFD on a day-to-day basis? | Can you describe the actual experience of living with CFD on a day-to-day basis? | |
| | Can you describe how you learnt to manage your CFD? | Can you describe how you learnt to manage your CFD? | | Can you describe how you learnt to manage your CFD? | Can you describe how you learnt to manage your CFD? |
| | Can you describe what it means to you having CFD? | Can you describe what it means to you having CFD? | Can you describe what it means to you having CFD? | Can you describe what it means to you having CFD? | |
| | Please discuss how you feel you manage your CFD | Please discuss how you feel you manage your CFD | Please discuss how you feel you manage your CFD | Please discuss how you feel you manage your CFD? | |
| | Can you discuss what it is like for you managing your CF and CFD? | Can you discuss what it is like for you managing your CF and CFD? | Can you discuss what it is like for you managing your CF and CFD? | Can you discuss what it is like for you managing your CF and CFD? | |
| | Can you discuss how you see your future living with CFD? | Can you discuss how you see your future living with CFD? | Can you discuss how you see your future living with CFD? | Can you discuss how you see your future living with CFD? | |
| | Can you discuss any ideas that you have that will help people with CF manage their CFD? | | | | Can you discuss any ideas that you have that will help people with CF manage their CFD? |
| prompts | | | | | |
| | Thought/feelings/typical day/ treatments/ | Compare and contrast to CF/ typical day/ images/ metaphors/ associations/ identity/thoughts/feelings/ | Impact on CF and vice versa/ thoughts/ feelings | Helps/hinders/ compare and contrast to CF/ control/ treatments | types resources/ education/ who to educate/support/what helps/ what doesn't help |

# Appendix 13: Initial noting and developing emergent themes



| Emergent themes | | transcript | Exploratory comments |
|---|---|---|---|
| | 1 | know putting in some extra Actrapid at the time. So, a different whole different | BD insulin to basal/prandially |
| | 2 | system to what I'm on now in terms of insulin erm yeah and that was it. | |
| | 3 | I: So, I know you said there was a lot going on at that time do you remember | |
| | 4 | how you coped when you said you just got on with it I mean... | |
| Coping w adversity | 5 | IV: Yeah cos it was my GCSE's as well which erm yeah it was all a bit crazy I lost a | bit crazy. understands all that going on. |
| | 6 | lot of my vision so I had to have large print paper at school erm again I just I never I | medically lot going on but GCSE on top of it; vision |
| | 7 | just literally got on with it. I don't remember I think because it was also my GCSE's that | + stoical |
| | 8 | was a massive distraction that I had a lot of school work to do erm and I wanted good | - distraction from what going on medically |
| | 9 | grades which I actually came out with which was erm a well not a surprise but it was a | motivation/preoccupation = GCSE |
| | 10 | surprise given everything that had happened that year. Erm yeah it just kind of I don't | wanting to do well. |
| Getting on with life | 11 | know if I ever really processed it, it was just one of those things and you just kind of | just got on with it. |
| | 12 | role with the punches erm yeah. | |
| | 13 | I: So, let's think about your experience of living with diabetes on a day to day | |
| | 14 | now sort of what's a typical day for you managing your diabetes would you say? | |
| Unstability of BG levels/ influences of food | 15 | IV: Erm it can be quite erm up and down at the moment with the lack of appetite | → yoyo effect - impact of poor appetite + supplements |
| | 16 | and a lot of the supplements which are really sugary it can mean that the blood sugars | |
| | 17 | yoyo erm and I have to keep a much closer eye on them now than I did a few years | → more monitoring now/than few years ago |
| | 18 | ago I think erm yeah, I do and the diabetes has a greater impact I think on my CF | greater impact on CF |
| | 19 | wellbeing. So erm if I'm having, I don't know any, yeah, it's if the more out of control | |
| Impact of CFRD control | 20 | they are the worse my CF symptoms will be I think so. | |
| | 21 | I: And by symptoms what do you mean? | |
| | 22 | IV: Coughing erm cough yeah coughing erm tiredness even erm appetite and erm | effect on CF symptoms |
| Impact of CFRD control | 23 | if I, if they do go out of control it seems to go it I might start coughing erm blood so it | pretty dramatic consequences high BG - coughing blood |
| | 24 | can have, it can have that kind of direct relation as well. | |
| | 25 | I: And do you see your diabetes has been sort of part of your CF or how would | |
| | 26 | you see it like in terms of so when your diabetes is bad your chest gets bad [yeah], | |
| | 27 | what relationship do you see? | |

| Emergent themes | | transcript | Exploratory comments |
|---|---|---|---|
| Separate conditions | 1 | IV: Erm I sort of have always seen them as quite separate probably erm...mmm I | Separate conditions |
| | 2 | don't really erm I don't know how to answer that question erm...yeah, I do, I don't | |
| | 3 | really know how to put the two together because yeah, that's not a good [laughs], | struggling + how see CFRD |
| | 4 | that's not an answer to the, what was the question again? | |
| | 5 | I: How do you see them being related to each other? | |
| relationship between diabetes and CF | 6 | IV: Yeah so, so yeah so, the worse my chest is the worse my diabetes is and the | direct relationship between diabetes + chest symptoms |
| | 7 | worse my diabetes is the worse my chest is so there is, there is that relationship. So, | |
| | 8 | but trying to keep them both under control is incredibly difficult. So erm so yeah so | control incredibly difficult - sense of |
| | 9 | there's always the ambition to keep it all under control but that's a lot easier said than | struggling with this |
| Unpredictability + management | 10 | done so I can have one day, an evening when I'll go out and sugars go all off and then | no patterns |
| | 11 | the next day partly because I'm exhausted anyway I'll be exhausted but then my chest | |
| | 12 | will be really bad and yeah. So erm so I have to do really good physios and things like | |
| | 13 | that and that's just from you know a few hours out erm so yeah so, the two are very | |
| | 14 | much directly related and just because just if my CF is good doesn't mean that my CF | |
| Unpredictability + management | 15 | is, if my chest is good doesn't mean my diabetes will be good though if that makes | - good chest doesn't necessarily mean well-controlled diabetes |
| | 16 | sense? And if my diabetes is good it doesn't mean my CF will be good it might just be | unpredictability of cf and diabetes |
| | 17 | easier to manage but I can have one bad and one good. Does that make any sense? | questioning - doesn't make sense to her? |
| | 18 | I: Yeah, I'm [laughs]... | looking for answers - sense making |
| | 19 | IV: Yeah, I don't know if that makes any sense at all but erm yeah. | lots of uncertainty |
| | 20 | I: And what's good diabetes mean to you? | |
| | 21 | IV: Less yoyoing, so less of the highs and then coming straight down to be low and | |
| | 22 | then you're having to have so you eat and then it seems to go up to like 15 but you've | |
| | 23 | had your insulin so almost immediately like an hour later you're like four so you have | |
| | 24 | to eat something and then it goes straight up and then up to like 14 and then you're | |
| | 25 | having insulin to bring it straight down and it just that can be exhausting and to keep | |
| | 26 | on top of that, that every time you eat it seems to go sky high but insulin it yeah it's | |
| unpredictability of CFRD | 27 | difficult to some days one dose of insulin will be enough and another day the insulin | difficulty of erratic nature of managing BG levels, eating + insulin |
| | 28 | dose will be completely different so yeah. That's what I find quite difficult is that it can | |
| | 29 | be quite erratic the erm yeah to if I can eat, if I eat the same things, do the same | |
| | 30 | things, have very similar days my insulin requirements can be quite different and I've | |

| Emergent themes | | transcript | Exploratory comments |
|---|---|---|---|
| looking for understand of relationship between CF + CFRD | 1 | always put that down to maybe something different is happening with my chest and | impact of CF |
| | 2 | that's causing you know slightly different, different readings so that's why it can be | |
| | 3 | quite difficult to keep on top of as well erm because it isn't just as straight forward as | not easy to manage sense of unpredictability + lack knowing |
| understanding insulin dosing | 4 | have the same doses all the time so yeah. | |
| | 5 | I: So that's a barrier really [~~12.57~~]. | |
| | 6 | IV: Yeah, yeah. | |
| | 7 | I: And what helps you manage your diabetes do you think? | |
| Role of blood glucose monitoring | 8 | IV: Erm what helps? Checking more frequently, blood sugars erm which isn't | checking BG helps but not always possible |
| | 9 | always easy I did just have a libre on I thought that was brilliant [laughs] I loved it and | help of new technology |
| Benefits of new technology | 10 | just through the ease of it and also seeing my patterns what was going on which I | |
| | 11 | hadn't know erm and the fact that it tells you whether you're going high or low rather | |
| | 12 | than just a figure. Erm so I did find that easier but beyond that erm...erm yeah | |
| influence of food | 13 | checking more and erm trying to eat, obviously eating regularly helps but that can be | difficult struggle balance between eating + BG when appetite poor |
| | 14 | quite difficult with erm poor appetite so yeah. | |
| | 15 | I: And can you tell me a bit about sort of your appetite and why it's difficult and | |
| | 16 | how that sort of all effects everything at the moment? | |
| | 17 | IV: Yeah so I don't really know why I've always had up until my mid to late 20's a | struggling with appetite for 4 years uncertain why went |
| managing consequences of CF | 18 | brilliant appetite erm and then about four years ago it just went pretty much | |
| | 19 | overnight erm I've never known why it went I also started vomiting a lot erm every | vicious circle eating, vomiting, chest worsening |
| | 20 | time I ate erm and then that would get worse if my chest was worse so erm so it's | |
| | 21 | been very difficult erm yeah I couldn't tell you what caused it, what was going on at | |
| | 22 | the time erm and its just never come back and that was about four years ago now. So | uncertainty |
| | 23 | erm its, which is really long, a lot longer than I erm I think it's been going on for so | |
| | 24 | yeah. | |
| | 25 | I: And how do you feel **** about having diabetes? | |
| | 26 | IV: Erm it's a bit of a pain, I do think that my CF would probably be going a little | pain |
| impact on CF | 27 | bit more straight forward if I didn't have it erm so yeah so it is erm a bit yeah it is | CF easier to manage without |
| | 28 | annoying but erm that's mostly it's just annoying. lots of hesitation | annoying - hesitation |
| CFRD is annoying | | 6 | |

| Emergent themes | | transcript | Exploratory comments |
|---|---|---|---|
| | 1 | I: Annoying. | |
| | 2 | IV: It's just yeah, annoying [laughs]. | inappropriate laugh |
| | 3 | I: And how did you learn to manage your diabetes? | |
| learning from a bad example | 4 | IV: Erm mmm partly probably through watching my sister actually learning what | learning from a bad example |
| | 5 | not to do cos she went through a phase when she was a teenager which she became | |
| | 6 | quite resentful cos she was diagnosed when she was eight and she became quite | |
| | 7 | resentful of it I think a bit bitter about it as in she wouldn't do her insulin, so she | |
| | 8 | needed a couple of ambulances out at the time in her late teens, she's now 35 but erm | |
| | 9 | and I don't, I couldn't understand why you would put yourself in that position to be | looking for a sense of understanding from her sisters position |
| | 10 | that vulnerable and so I think that's seeing that made me realise what not to do but | |
| | 11 | also I think erm well yeah that was that's probably the main thing yeah. | |
| | 12 | I: And how did you get your skills to manage your diabetes on a day to day? I | |
| | 13 | mean were they from your sister or were or...? | |
| | 14 | IV: Mmm...oh god, I could say myself, I didn't really have much input at the time | modality - admitting skills from herself |
| lack of professional support | 15 | when I was diagnosed erm at the time I was seeing a CF team locally to where I live in | |
| | 16 | [redacted] erm and I yeah, I didn't really have much input from anyone really. So, it was | sense of being on own - not much input |
| | 17 | mostly erm my family, my mum was brilliant erm she'd been having to do my sisters | support from family |
| support from family | 18 | injections from my sister being eight so erm, so yeah probably more from my mum | |
| | 19 | actually erm I came yeah. | |
| | 20 | I: And erm sort of you said your sisters got diabetes and you see her sort of as a | |
| | 21 | way to sort of not manage your diabetes and that's how you knew then [mmm yeah]. | |
| | 22 | Is I mean and your mum was a good support, was there – do you feel that you needed | |
| | 23 | more support at that time or was the level of support okay **** and what would you | |
| | 24 | say about the support when you were diagnosed? | |
| | | (long pause) | |
| | 25 | IV: Mmm erm.....the level of support erm it was okay, it wasn't I think if I hadn't | hesitant about amount of support from team - sense of not wanting to admit wasn't good |
| | 26 | had the support of my family and particularly my mum and things I would have found | |
| | 27 | it poor erm because there was no one to talk to, my consultant at the time wasn't | |
| lack of professional support | 28 | great and he would always mix me and my sister up anyway erm so its yeah it wasn't | |
| | 29 | great. And my consultant wasn't someone I felt that I could talk to particularly openly | couldn't talk to consultant - |

| Emergent themes | | transcript | Exploratory comments |
|---|---|---|---|
| | 1 | and the other staff again it was a bit rubbish but as I said that was, that wasn't here | local support rubbish |
| | 2 | that was in [redacted] erm and then... | |
| | 3 | I: And 15 years ago, you say was that right? | |
| Role of specialist team | 4 | IV: Yes, so that was in 2000, yeah. Erm yeah and then I think because it was CF | 17 years ago |
| | 5 | related diabetes the [redacted] felt that it would be better if I came here, started | Specialist Centres took over care the |
| | 6 | coming here but erm even then all my notes never seemed to transfer across so I've | but inadequacies in records/transfer, |
| | 7 | had a lot of people here question when I was diagnosed, thinking it was recent when | at the time. |
| | 8 | actually it was 17 years ago and stuff like that erm that was particularly when I first | |
| | 9 | moved across, I'm not too sure about now erm and erm yeah, yeah. | |
| | 10 | I: And erm how do you feel that you manage your diabetes now? How... | |
| role of blood glucose testing | 11 | IV: I think I manage it okay I know I could do better erm I do check my blood | Sees the negatives |
| | 12 | probably about four times a day but I know I would do better if I was doing it more | BG-testing feels needs to improve on |
| | 13 | particularly after meals, I'm good at doing it before meals but not so good at doing it | than |
| | 14 | after meals. Erm and that's probably, possibly why I do yoyo because I don't know | trying to make sense of yoyo |
| Trying to make sense of BG levels | 15 | what's happening after I've eaten whether it's been too much or too little or whatever | |
| | 16 | erm yeah so, I do think it's okay but if definitely could be better. | |
| | 17 | I: What would make it better do you think other than... | |
| | 18 | IV: Testing, yeah testing my blood more erm.....mmm...I think that's mostly, | long pause - sees testing as |
| role of blood glucose testing | 19 | mostly it is the te, is having more readings to go by erm yeah but it's very inconvenient | key to making CARD better managed |
| | 20 | to be checking all the time you know when you're out and about and you have to get | |
| | 21 | your finger pricker out and yeah it's you can't really do it well I don't feel comfortable | visibility of BG checking - not |
| | 22 | doing it in front of strangers erm you know it's the same with insulin if you're out and | comfortable in public (+ insulin also) |
| Impacts of treatments and monitoring | 23 | about some people will just do it in front of people I'm much less comfortable doing | |
| | 24 | that I'd rather take myself off to a loo or something private and if that's not accessible | |
| | 25 | then I will wait until I can get to a place where I can do it privately which isn't great | wanting to do insulin if not a |
| food | 26 | because that's when erm if I've eaten then I have to wait before I do my insulin and | 'private place' - acknowledges negative |
| | 27 | stuff like that. So, if I'm out and about that can be, that causes issues so if I were more | effect on CARD. |
| | 28 | comfortable doing all of these things in front of people that would also help but I don't | laughs |
| | 29 | know that that's gonna change any time soon yeah. | - don't feel will become comfortable |
| | | | doing treatment / monitoring in front people |
| | | | [had dm for 17 years - unlikely |
| | | | to change now.? |

# Appendix 14: Illustrations of emergent themes

| cluster label - MAGIC 02 | theme | Page & number | quote/keyword |
|---|---|---|---|
| | managing the diabetes diagnosis | 3,17 | So, there was so much going on in the space of about six months that erm it just kind of was another thing that came |
| | | 3,28 | it was just constant erm finger pricks I found that actually harder than the insulin injections [laughs] |
| | | 4,5 | Yeah cos it was my GCSE's as well which erm yeah it was all a bit crazy I lost a lot of my vision so I had to have large print paper at school erm again I just I never I just literally got on with it |
| | | 4,11 | I don't know if I ever really processed it, it was just one of those things and you just kind of role with the punches erm yeah |
| | | | |
| **learning to live with diabetes** | managing the impact of CF and its treatments | 3,4 | with all of the yoyoing of the steroids then my glucose levels went all over the place so I got diabetes |
| | | 3,5 | huge doses of steroids for at least a year erm when I was 14 to 15 |
| | | 3,10 | I think because I'd been having to watch my glucose levels for several weeks because of the steroids I think it was at the back of my mind |
| | | 6,17 | I've always had up until my mid to late 20's a brilliant appetite erm and then about four years ago it just went pretty much overnight erm I've never known why it went I also started vomiting a lot |
| | | 6,20 | that would get worse if my chest was worse so erm so it's been very difficult |
| | | 6,26 | Erm it's a bit of a pain |

| | | | |
|---|---|---|---|
| | | 8,18 | I don't feel comfortable doing it in front of strangers erm you know it's the same with insulin if you're out and about some people will just do it in front of people I'm much less comfortable doing that |
| | | 8,27 | So, if I'm out and about that can be, that causes issues |
| | | 18,3 | I'm having to have such high sugar supplements I think it was so much better before I had to rely on those |
| | | 22,10 | steroids you know you're doing that's actually something beneficial for your CF but it then throws you, all your diabetes into yeah it goes haywire totally |
| | | 22,13 | always difficult to then especially with a constantly changing dose of steroids to keep on top of that yeah as well as all the other things |
| | | | |
| | influence of family members with diabetes | 3,19 | also my older sister has it so I knew about it |
| | | 3,20 | I knew what she had to do erm in terms of blood sugars and things so it wasn't a whole new learning curve for me |
| | | 3,21 | so erm so it wasn't maybe as scary to me |
| | | 7,4 | Erm mmm partly probably through watching my sister actually learning what not to do cos |
| | | 7,9 | I couldn't understand why you would put yourself in that position to be that vulnerable and so I think that's seeing that made me realise what not to do |
| | | 20,10 | I definitely wasn't as scared of it because I'd been watching her do it for the you know for the previous 10 years |
| | | | |
| | who provided CFD support | 7,14 | I didn't really have much input at the time when I was diagnosed |
| | | 7,16 | So, it was mostly erm my family, my mum was brilliant |
| | | 7,25 | Mmm erm……the level of support erm it was okay, it wasn't I think if I hadn't had the support of my family and particularly my mum and things I would have found it poor erm because there was no one to talk to, my consultant at the time wasn't great |

| | | | |
|---|---|---|---|
| | | 7,29 | And my consultant wasn't someone I felt that I could talk to particularly openly and the other staff again it was a bit rubbish |
| | | 8,4 | then I think because it was CF related diabetes the ***** felt that it would be better if I came here, started coming here |
| | | 10,14 | I don't think I've ever been given anything |
| | | 10,16 | [laughs] Erm is there additional help? Like yeah, it's never been erm offered to me I wouldn't have thought there was other ways of doing things |
| | | 10,18 | I didn't know there erm if there is additional help I definitely don't know about it so. |
| | | 13,3 | I don't, I tend not to like the CF forum cos I actually find it quite depressing |
| | | 13,27 | there's a lot of people, you get some people who are really into it but they might be further along in their condition than you and they talk about things that are potentially in your future |
| | | 14,1 | I'll deal it with it when I get there, |
| | | 14,20 | I make sure I stay out of everyone's way. So, there's definitely not a way for me to meet people unless it would be online and as I avoid all of that anyway I kind of no I don't have any peer |
| | | 14,25 | I think a lot of people would get a lot out of that |
| | | 14,28 | So, I think yes that would be good, may, not necessarily for me but I don't, things could change if things get more difficult maybe I would rely on those things more peer support |
| | | 15,12 | erm myself again blogs, yes erm I will see blogs I don't necessarily follow medical blogs |
| | | 15,14 | I just go, dip in and out if and when I want to know something or find out something |
| | | 15,15 | So yes, I would rely on them blogs and what other people say when I'm trying to get more information |
| | | 18,18 | I do find the appointments quite difficult because I feel like I live here these days and the diabetes appointments tend to be very short |
| | | 18,22 | so it often feels like I've come in for nothing because a lot of the time nothing changes |
| | | 18,24 | So, it does feel like it's a lot of effort to have to, to do the diabetes appointments as well that is yeah. |

| | | | |
|---|---|---|---|
| | | 9,19 | Erm that would be nice if the two could be combined |
| | | | |
| | | | |
| | diabetes treatment draws attention from others | 9,3 | Erm I don't like it just because I think other people get scared |
| | | 9,4 | 'Oh what's wrong?' as if something's happening and then they start to panic so I'm trying not to panic other people |
| | | | |
| | words to describe CFD | 6,28 | mostly it's just annoying |
| | | 20,20 | Mmm it's a pain [laughs]. |
| | | 21,1 | it's a pain it gets in the way |
| | | 21,1 | its nuisance having to check or erm go off to do insulin or you're not always in a or you're at work and you start to feel like you're going low but you're in a meeting and it's just a nuisance it's you know |
| | | | |
| **An unpredictable world** | managing changes in health | 1,9 | Okay erm at the moment it's getting harder erm that's just within the last couple of years it kind of has got a lot harder very quickly |
| | | 2,12 | its limiting me quite a lot. |
| | | 23,25 | because I'm on steroids and supplements which effect my diabetes a lot more the diabetes has got a bigger role now so |
| | | | |
| | looking for an understanding of the relationship between CF and CFD | 4,18 | the diabetes has a greater impact I think on my CF wellbeing |
| | | 5,1 | Erm I sort of have always seen them as quite separate probably erm |

| | | | |
|---|---|---|---|
| | | 5,6 | the worse my chest is the worse my diabetes is and the worse my diabetes is the worse my chest is so there is, there is that relationship |
| | | 5,15 | if my chest is good doesn't mean my diabetes will be good though if that makes sense? And if my diabetes is good it doesn't mean my CF will be good it might just be easier to manage but I can have one bad and one good. |
| | | 5,29 | if I eat the same things, do the same things, have very similar days, my insulin requirements can be quite different and I've always put that down to maybe something different is happening with my chest |
| | | 6,26 | I do think that my CF would probably be going a little bit more straight forward if I didn't have it |
| | | 23,1 | , I think I've always seen them as separate and that's possibly because my sisters got diabetes so I see it as a separate condition but I think it probably should be seen as one and treating both together |
| | | 23,9 | even in clinic the two are separated very much |
| | | | |
| | unpredictability managing CF and CFD | 4,15 | Erm it can be quite erm up and down at the moment with the lack of appetite and a lot of the supplements which are really sugary it can mean that the blood sugars yoyo |
| | | 5,9 | there's always the ambition to keep it all under control but that's a lot easier said than done so I can have one day |
| | | 5,25 | and it just that can be exhausting and to keep on top of that, that every time you eat it seems to go sky high but insulin it yeah it's difficult to some days one dose of insulin will be enough and another day the insulin dose will be completely different so yeah |
| | | 5,29 | That's what I find quite difficult is that it can be quite erratic |
| | | 24,19 | you do feel out of control a lot of the time with diabetes cos you think you're doing the right thing and you can be checking and it's still the wrong thing and that can be frustrating |
| | | | |
| | differences to other types of diabetes | 11,18 | what I've been told is that, that they're very different |

| | | | |
|---|---|---|---|
| | | 11,22 | Erm so again I don't know why or how that's not really been erm if it was explained to me |
| | | | |
| | | | |
| | impact of CFD control | 4,17 | I have to keep a much closer eye on them now than I did a few years ago |
| | | 4,19 | it's if the more out of control they are the worse my CF symptoms will be I think so |
| | | 4,23 | , if they do go out of control it seems to go it I might start coughing erm blood |
| | | 5,8 | but trying to keep them both under control is incredibly difficult |
| | | 8,13 | I think I manage it okay I know I could do better erm |
| | | 12,2 | I've only worked out through myself that if my chest is bad that my blood sugars will be more erratic |
| | | | |
| | influence of the individual | 3,18 | you just deal with |
| | | 3,29 | but I got over that relatively quickly erm |
| | | 8,27 | I were more comfortable doing all of these things in front of people that would also help but I don't know that that's gonna change any time soon yeah |
| | | 9,10 | it was a need to know basis |
| | | 9,15 | it's definitely not something I'm erm loud, loud and proud about but yeah. |
| | | 10,21 | Erm I kind of tend to be just get on with it I tend not to fuss or ask questions so I don't know how much I would take up additional help erm |
| | | 11,1 | definitely just because of my personality erm I tend to have to be pushed if I'm feeling unwell just to call the CF nurses |
| | | 11,9 | I'll be determined that I can do it myself |
| | | 13,14 | I think because I'm just far too private maybe that I don't like putting things out there, |
| | | 14,5 | I'd rather try and keep in higher spirits if I can you know |
| | | 14,11 | some people who dwell on it and its and moan and I just no, I don't have time for that |
| | | 24,21 | my personality I need to be in control [laughs] erm so I, that is erm difficult and so then when I am losing control that can be incredibly erm frustrating erm |

| | | | |
|---|---|---|---|
| | | 25,24 | Me and then my condition [yeah] is always very much just a part of it but not who I was |
| | | 25,27 | whereas for me it was just I have to do these things but there's oth, a load of other things I want, I enjoy getting on with so |
| | | | |
| | The future with CF and CFD | 6,9 | I did just have a libre on I thought that was brilliant [laughs] I loved it and just through the ease of it and also seeing my patterns what was going on |
| | | 9,20 | I think I get nervous about my eye sight erm I would hate for that to go |
| | | 9,22 | I think that is definitely a worry of mine particularly if they yoyo and I'm on a drug at the moment for my vasculitis |
| | | 10,1 | when I think of my conditions CF is definitely the main one I don't think CF and diabetes, |
| | | 10,3 | So, I do see them separately so when I think about my future it def, it is more of a CF dominated future than a diabetes one |
| | | 10,6 | the CF future which is now more prominent in my head |
| | | | |
| **skills needed to self-manage CFD** | understanding blood glucose monitoring | 8,6 | Erm what helps? Checking more frequently, blood sugars erm which isn't always easy |
| | | 6,13 | checking more and erm trying to eat, obviously eating regularly helps but that can be quite difficult with erm poor appetite so yeah. |
| | | 8,11 | I do check my blood probably about four times a day but I know I would do better if I was doing it more particularly after meals |
| | | 8,14 | Erm and that's probably, possibly why I do yoyo because I don't know what's happening after I've eaten |
| | | 8,18 | testing my blood more erm……mmm…I think that's mostly, mostly it is the te, is having more readings to go by erm yeah but it's very inconvenient to be checking all the time you know when you're out and about |
| | | 16,24 | it's not rare it's something I'm doing constantly it should be something I should be able to manage better |

| | | | |
|---|---|---|---|
| | | 17,1 | cos I don't know what I'm doing wrong for it to go so high and to stop the yoyoing. |
| | | 17,8 | it's not just going up a little bit and coming down a little bit it goes really high and then comes down |
| | | | |
| | | | |
| | understanding insulin | 6,3 | it isn't just as straight forward as have the same doses all the time so yeah |
| | | 8,21 | I don't feel comfortable doing it in front of strangers erm you know it's the same with insulin |
| | | 16,12 | so I seem to be having the right amount of insulin to bring me back down but how do you stop going so high in the first place? |
| | | 17,14 | Erm oh dear it's probably largely guess work …. |
| | | | |
| | managing food | 4,15 | Erm it can be quite erm up and down at the moment with the lack of appetite and a lot of the supplements which are really sugary it can mean that the blood sugars yoyo |
| | | 12,10 | how to calculate how much insulin to have with erm a meal in terms of carbohydrates in grams. Erm so that actually was helpful |
| | | 12,14 | didn't realise that there was a link between how much insulin you have with the carbohydrate |
| | | 17,7 | its mostly that it's the because I would have great levels if I didn't eat [laughs] and it's the eating and things that seem to throw them into chaos |
| | | | |
| | self-directed learning | 7,14 | Mmm…oh god, I could say myself, |
| | | 17,18 | I've got there eventually to the right levels a |
| | | | |
| **what information and support is needed** | lack of knowledge | 11,26 | Erm so again I don't know why or how that's not really been erm if it was explained to me |

| | | | |
|---|---|---|---|
| | | 16,14 | after 17 years I still don't know which is actually probably pretty terrible erm I should know these things and I don't. |
| | | 17,11 | how, I don't know how to stop, how to stop it going so high yeah. |
| | | | |
| | format of support | 13,7 | I would use an online, online is a way that I would try and find things out yeah |
| | | 13,10 | I'll just Google the question a lot of the time and then see what comes up. |
| | | 13,15 | I'll see if someone else is going through it and if they're not then I'm not going to fuss about starting it |
| | | 13,17 | I think that's probably also why I like online because it means that I don't have to do group activities necessarily |
| | | 13,20 | I went to it but I had to go to a group session and I never went back cos it was so depressing |
| | | 13,24 | you get a lot of people who are very open but can be quite erm down and down about things and I'm just not interested in that |
| | | 15,24 | It kind of depends what the leaflets are about I guess |
| | | 16,3 | but again it has to be something maybe that would change my care in some way or revolutionise it, |
| | | | |
| | what information is needed | 11,17 | Erm yeah there's very little CF diabetes information if I have looked its more just diabetes |
| | | 11,27 | , I would quite like to know why it's different actually erm cos I don't know if that would help me manage my diabetes better if I know what to expect or why there are certain triggers or what potential, what the potential triggers are I mean I have no idea |
| | | 16,10 | Erm mmm how to manage meal times cos I still don't think I have that down because I still when I eat seem to go up really high |
| | | 17,23 | I think all information helps |
| | | 17, 23 | I never do ketone checks. I don't know if I'm supposed to |
| | | 17,24 | I think all information is good I don't think there's anything that I wouldn't not want to know |
| | | | |

| | | | |
|---|---|---|---|
| | demands on time | 12,18 | so if there are going to be things like that to help you manage better I think a week off work is way too much |
| | | 12,21 | when you're having to take so much time off for appointments, so much time off for admissions or for IV's then you've got your holidays on top |
| | | 12,24 | you really want the holiday leave |
| | | 12,26 | yeah it needs to be short manageable things or maybe online |
| | | 19,16 | but it definitely feels like its once every couple of weeks. |
| | | 19,23 | when it just for 20 minutes you think oh god I've travelled for three hours just for 20 minutes so yeah |
| | | | |

# Appendix 15: Emergent themes- searching for connections

Similar emergent themes were merged together (participant 2)

| **similarities** | **merging similar themes** |
|---|---|
| impact of health changes | managing changes in health |
| changes in health | |
| | |
| influence of family members with diabetes | influence of family members with diabetes |
| | |
| education regarding food and insulin | understanding insulin |
| judging insulin doses | |
| | |
| influence of food | |
| role of food | managing food |
| managing food | |
| | |
| instability of blood glucose levels | |
| role of blood glucose monitoring | understanding blood glucose monitoring |
| role of blood glucose testing | |
| trying to make sense of blood glucose levels | |
| variability/unpredictability of blood glucose levels | |
| | |
| lack of professional support | |
| support from family | who provided CFD support |
| role of specialist team | |
| uncertainty re support | |
| role of peer support | |
| peer support via blog | |
| use of CF forums | |
| dislikes of groups | |
| negatives of forums | |
| value of CFD clinic | |
| | |
| coping in adversity | |
| getting on with life | |
| disclosure | |
| attitude/ personal attributes | Influence of the individual |
| personal attributes | |
| personality | |
| positive attitudes | |

| | |
|---|---|
| positive attitudes | |
| CF and CFD just something to manage | |
| | |
| words to describe CFD | words to describe CFD |
| an additional problem | |
| CFD is annoying | |
| CFD is a pain | |
| | |
| demands on time | demands on time |
| limited demands on time | |
| | |
| separate conditions | |
| relationships between diabetes and CF | looking for an understanding of the relationship between CF and CFD |
| looking for an understanding of the relationship between CF and CFD | |
| increase knowledge of link between CF and CFD | |
| | |
| unpredictability in management | unpredictability managing CF and CFD |
| unpredictability of CFD | |
| | |
| impact of CF treatments | managing the impact of CF and its treatments |
| impact of CF | |
| impact of treatments and monitoring | |
| impact of steroids | |
| managing consequences of CF | |
| treatment impacting on diabetes | |
| | |
| impact of control | |
| impact of CFD control | impact of CFD control |
| | |
| complications of diabetes | The future with CF and CFD |
| impact of CF on future | |
| benefits of new technology | |
| | |
| use of leaflets | role of leaflets |
| role of leaflets | |
| | |
| self-directed learning | self-directed learning |
| learning through doing | |
| | |
| lack of CFD information | |
| what information is needed | what information is needed |

# Appendix 16: Clustering themes

| cluster label - MAGIC 02 | theme |
|---|---|
| | managing the diabetes diagnosis |
| **learning to live with diabetes** | managing the impact of CF and its treatments |
| | influence of family members with diabetes |
| | who provided CFD support |
| | |
| | diabetes treatment draws attention from others |
| | words to describe CFD |
| **An unpredictable world** | managing changes in health |
| | looking for an understanding of the relationship between CF and CFD |
| | unpredictability managing CF and CFD |
| | differences to other types of diabetes |
| | impact of CFD control |
| | influence of the individual |
| | The future with CF and CFD |
| **skills needed to self-manage CFD** | understanding blood glucose monitoring |
| | understanding insulin |
| | managing food |
| | self-directed learning |
| **what information and support is needed** | lack of knowledge |
| | format of support |
| | what information is needed |
| | demands on time |

# Appendix 17 - Cross- case analysis- looking for connections between identified themes

| cluster label - MAGIC 01 | theme MAGIC 01 | cluster label - MAGIC 02 | theme MAGIC 02 | cluster label - MAGIC 03 | theme MAGIC 03 |
|---|---|---|---|---|---|
| | response to CFD diagnosis | | managing the diabetes diagnosis | impact of CFD | diagnosis |
| putting CFD in perspective | prioritising health concerns and management | learning to live with diabetes | managing the impact of CF and its treatments | | day-to-day life |
| | achieving a balance | | influence of family members with diabetes | | health |
| | | | who provided CFD support | | future |
| | CF and CFD - unpredictability and variability | | | | mental demands of trying to manage CFD |
| | part of CF routine | | diabetes treatment draws attention from others | key self-management activities in self-manage CFD | managing blood glucose levels |
| | | | words to describe CFD | | Understanding insulin |
| essentail skills required for self-management | BG monitoring | An unpredicatable world | managing changes in health | | managing food |
| | managing diet | | looking for an understanding of the relationship between CF and CFD | | role of exercise |
| | insulin the unknown condrum | | unpredictability managing CF and CFD | How should support be provided and by what means | How should support be provided and by what means |
| | experiential learning | | differences to other types of diabetes | | |
| | influence of activity | | impact of CFD control | | |
| | knowledge needs | | influence of the individual | | |
| | | | The future with CF and CFD | | |
| what makes CFD difficult to self-manage | managing treatments | skills needs to self-manage CFD | understanding blood glucose monitoring | | |
| | support | | understanding insulin | | |
| | self | | managing food | | |
| what helps CFD self-management | managing treatments | | self-directed learning | | |
| | significance of control | what information and support is needed | lack of knowledge | | |
| | self | | format of support | | |
| | uniqueness of CFD | | what information is needed | | |
| | | | demands on time | | |
| specific CFD management demands need to be addressed | role of peers | | | | |
| | practical vs medical advice | | | | |
| | resource and support requirements for self-management | | | | |

| cluster label 04 | theme MAGIC 04 | cluster label 05 | theme MAGIC 05 | cluster label MAGIC 06 | theme MAGIC 06 |
|---|---|---|---|---|---|
| | managing the diagnosis | | the diagnosis of CFD | | managing the diagnosis of CFD |
| **Adjusting to living with CFD** | | **adjusting to living with CFD** | learning to live with CFD | **putting CFD into perspective** | learning to live with CFD |
| | impact of CFD | | relationship between CF and CFD | | impact of personal attributes |
| **Achieving a balance between CF and CFD** | impact of CF treatments on CFD (a spiral of deterioration) | | future with CFD | | living with the variability and unpredictability |
| | relationship between CF and CFD | | | | relationship between CF and CFD |
| **How to self-manage CFD** | role of diet | **What is needed to manage CFD** | managing blood glucose levels | | barriers and facilitators to CFD management |
| | Managing insulin therapy | | management of insulin | | the future |
| | managing blood glucose levels | | influence of exercise | | |
| | impact of exercise | | Dietary management | | |
| **Need for information and support** | what information | **What information and support is neede and how should it be delivered** | | **Essential self-management skills** | role of exercise |
| | how to provide | | what information is needed | | role of diet |
| | | | how should support be provided | | role of blood glucose monitoring |
| | | | | | insulin |
| | | | | | |
| | | | | **Need for information and support** | what information is needed |
| | | | | | How should support be provided |
| | | | | | |
| | | | | | |

| cluster label MAGIC 07 | theme MAGIC 07 | cluster label Magic 08 | theme Magic 08 |
|---|---|---|---|
| reaching a position of acceptance with CFD | **managing the diagnosis of CFD** | | managing the diagnosis of CFD |
| | **impact** | | |
| | **identity** | managing the impact of CFD | learning to manage CFD |
| | **relationship with** | | what it means to have CFD |
| | **future** | | relationship between CF and CFD |
| learning to mange CFD | **challenges** | | disclosure |
| | **barriers** | | the future |
| | **insulin the unknown conundrum** | | |
| | **diet** | **living with CFD on a day to day basis** | role of blood glucose testing |
| | **personal attributes** | | managing insulin injections |
| balancing act between managing CF and CFD | **balancing act between managing CF and CFD** | | managing nutritional status |
| What are the requirements for information and support | **what information is needed** | | impact of exercise |
| | **what support is needed and by whom** | | |
| | **How should information be provided** | **the unmet need for information and support** | how should support be delivered to people with CFD |
| | | | what information is needed |
| | | | |
| | | | |
| | | | |

# Appendix 18: Cross- case analysis – merge identified themes to develop superordinate themes

| Master themes | theme MAGIC 01 | theme MAGIC 02 | theme MAGIC 03 | theme MAGIC 04 | theme MAGIC 05 | theme MAGIC 06 | theme MAGIC 07 | theme Magic 08 |
|---|---|---|---|---|---|---|---|---|
| | response to CFD diagnosis | managing the diabetes diagnosis | diagnosis | managing the diagnosis | the diagnosis of CFD | managing the diagnosis of CFD | managing the diagnosis of CFD | managing the diagnosis of CFD |
| forming a realtionship with CFD | learning to live with CFD | learning to live with CFD | day-to-day life | learning to live with CFD | learning to live with CFD | learning to live with CFD | learning to live with CFD | learning to manage CFD |
| | CF and CFD - unpredictability and variability | looking for an understanding of the relationship between CF | | relationship between CF and CFD | relationship between CF and CFD | relationship between CF and CFD | balancing act between managing CF and CFD | relationship between CF and CFD |
| | | The future with CF and | future | | future with CFD | the future | future | the future |
| Balancing the CFD self-managemnt triad | insulin the unknown condrum | understanding insulin | Understanding insulin | Managing insulin therapy | management of insulin | managing insulin | insulin the unknown conundrum | managing insulin injections |
| | BG monitoring | understanding blood glucose monitoring | managing blood glucose levels | managing blood glucose levels | managing blood glucose levels | role of blood glucose monitoring | barriers to blood glucose monitoring | role of blood glucose testing |
| | managing diet | managing food | managing food | role of diet | Dietary management | role of diet | diet | managing nutritional status |
| | influence of activity | | role of exercise | impact of exercise | influence of exercise | role of exercise | | impact of exercise |
| | self | influence of the individual | | | | impact of personal attributes | personal attributes | |
| | prioritising health concerns and management | managing changes in health | health | | | | | |
| The unmet need for information and support | resource and support requirements for self-management | what information is needed | How should support be provided and by what means | what information | what information is needed | what information is needed | what information is needed | what information is needed |
| | | format of support | | how to provide | how should support be provided | How should support be provided | what support is needed and by whom | how should support be delivered to people with |
| | | | | | | | How should information be provided | |
| | achieving a balance | managing the impact of CF and its treatments | mental demands of trying to manage CFD | impact of CFD | | living with the variability and unpredictability | impact | what it means to have CFD |
| | part of CF routine | influence of family members with diabetes | | impact of CF treatments on CFD (a spiral of | | barriers and facilitators to CFD management | identity | disclosure |
| | | who provided CFD support | | | | | relationship with | |
| | experiential learning | diabetes treatment draws attention from others | | | | | barriers | |
| | managing treatments | words to describe CFD | | | | | challenges | |
| | support | demands on time | | | | | | |
| | managing treatments | differences to other types of diabetes | | | | | | |
| | significance of control | impact of CFD control | | | | | | |
| | uniqueness of CFD | | | | | | | |
| | | self-directed learning | | | | | | |
| | knowledge needs | lack of knowledge | | | | | | |
| | role of peers | | | | | | | |
| | practical vs medical advice | | | | | | | |
| | | unpredictability managing CF and CFD | | | | | | |

**Appendix 19: MAGIC curriculum CFD beginner**


# Curriculum – CFD Beginner

| CFD BEGINNER | | |
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| **Learning goals** | **Content** | **Resources/ notes** |
| • **Be able to describe what CFD is** | **What is CFD?**<br>• Brief explanation diabetes caused by lack of insulin and resultant increase in blood glucose<br>• Discussion of delay and reduction in insulin-<br>• Who affects | • video – brief overview CFD<br>• module text |
| • **Know where insulin is produced**<br>• **Know why cells need glucose**<br>• **Understand how insulin lowers blood glucose levels**<br>• **Understand that normally insulin is produced automatically** | **Why do we need insulin?**<br>• Explanation of journey from carbohydrates in food we eat to energy for use by our bodies.<br>• Discussion what happens in someone without diabetes | • Module text<br>• Diagram food to energy<br>• Chart normal insulin production |
| • **Discuss what happens when the body does not produce enough insulin** | **What happens without insulin?**<br>• Discussion of consequences of not having enough insulin- increase glucose production by liver, breakdown of body's protein and fat stores and consequent weight loss, muscle weakness, tiredness. Spill over of glucose into urine and subsequent polyuria and polydipsia. | • Module text<br>• Diabetes UK what is diabetes video https://www.diabetes.org.uk/diabetes-the-basics/diabetes-and-the-body |

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| • **Identify why having CF causes CFD**<br>• **Be aware that CFD is not preventable** | **Why does CF cause diabetes?**<br>• Discussion of link between pancreatic disease in CF and development of CFD | • Module text |
| • **Be aware of the effects of CFD on CF** | **Relationship between CF and CFD**<br>• Brief discussion of consequences of uncontrolled CFD on lungs and nutrition | • Module text |
| • **Be able to identify the symptoms of CFD** | **Symptoms of CFD**<br>• Discussion of symptoms some people who develop CFD experienced | • Module text<br>• Personal log- my symptoms of CFD |
| • **Know why insulin needs to be injected**<br>• **Know the difference between long-acting and quick-acting insulin**<br>• **Be able to identify own insulin regimen**<br>• **Know what foods affect blood glucose levels**<br>• **Give examples of carbohydrate containing foods** | **How is CFD treated?**<br>• Insulin<br>  ○ How given<br>  ○ Types of insulin- long-acting and quick-acting<br>• Diet<br>  ○ Food broken down into three main nutrients- protein, fats and carbohydrates<br>  ○ What are carbohydrate containing foods<br>  ○ Limit sugary drinks, sweets and fruit juices to meals | • Module text<br>• Personal experience quotes<br>• Personal log- my insulin |

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|---|---|---|
| • **Know how to test blood glucose levels**<br>• **Understand why to test blood glucose levels**<br>• **Understand when to test blood glucose levels**<br>• **Know what normal blood glucose levels are for people without diabetes**<br>• **Demonstrate what ideal blood glucose levels are** | **Blood glucose**<br>• Why test<br>  ○ Will not know what blood glucose are by the way feel<br>  ○ Include testing to inform treatment and dietary decisions, high and low blood glucose levels, lifestyle choices<br>• How to test<br>  ○ Give stepwise instructions how to check blood glucose levels in video<br>  ○ Highlight use of side of fingertip rather than pad<br>• When to test<br>  ○ Discussion of when to test if on long-acting insulin and meaning of results<br>  ○ Discussion when to test if on quick-acting insulin and meaning of results<br>• What do they mean? | • Module text<br>• Video of how to use blood glucose monitor<br>• Personal log- my blood glucose monitoring |

| | | |
|---|---|---|
| | ○ What are normal blood glucose levels for people without diabetes<br>○ Discussion of initial blood glucose targets in the early days of managing CFD- 4.5-7.5 on waking and between meals, <10mmol 1½-2 hours after meals<br>○ Discussion that normal to take a while to get blood glucose levels under control | |
| • **Appreciate how other people manage with CFD** | **Managing your diagnosis**<br>• Explanation that normal to feel overwhelmed, experiencing managing CFD is challenging<br>• Team there to offer support | • Module text<br>• Videos and quotes – experiences of managing CFD diagnosis |
| • **To review knowledge and skills acquired through module** | **Review of module** | • Test your learning quiz<br>• Course moderator to contact participant 7 days after being given module, then at followed agreed timepoint to review questions, understanding and readiness for next module/ or more follow up sessions if required |